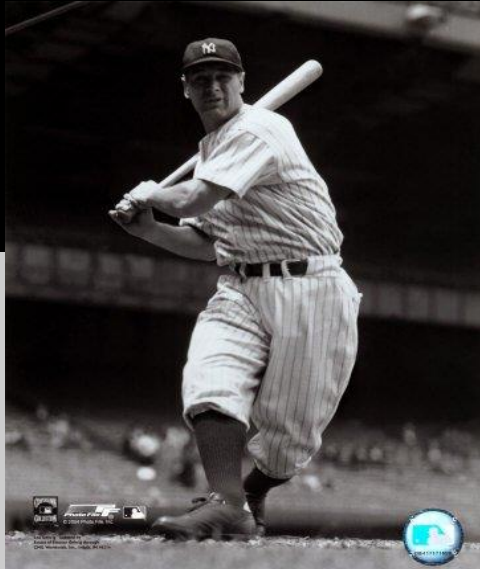


It's not just Lou's disease.....



..... It's everyone's!

# ALS Family Manual

## ALS Family Manual



**The Amyotrophic  
Lateral Sclerosis Association**

*St. Louis Regional Chapter*

Helping people.



United Way  
of Greater St. Louis

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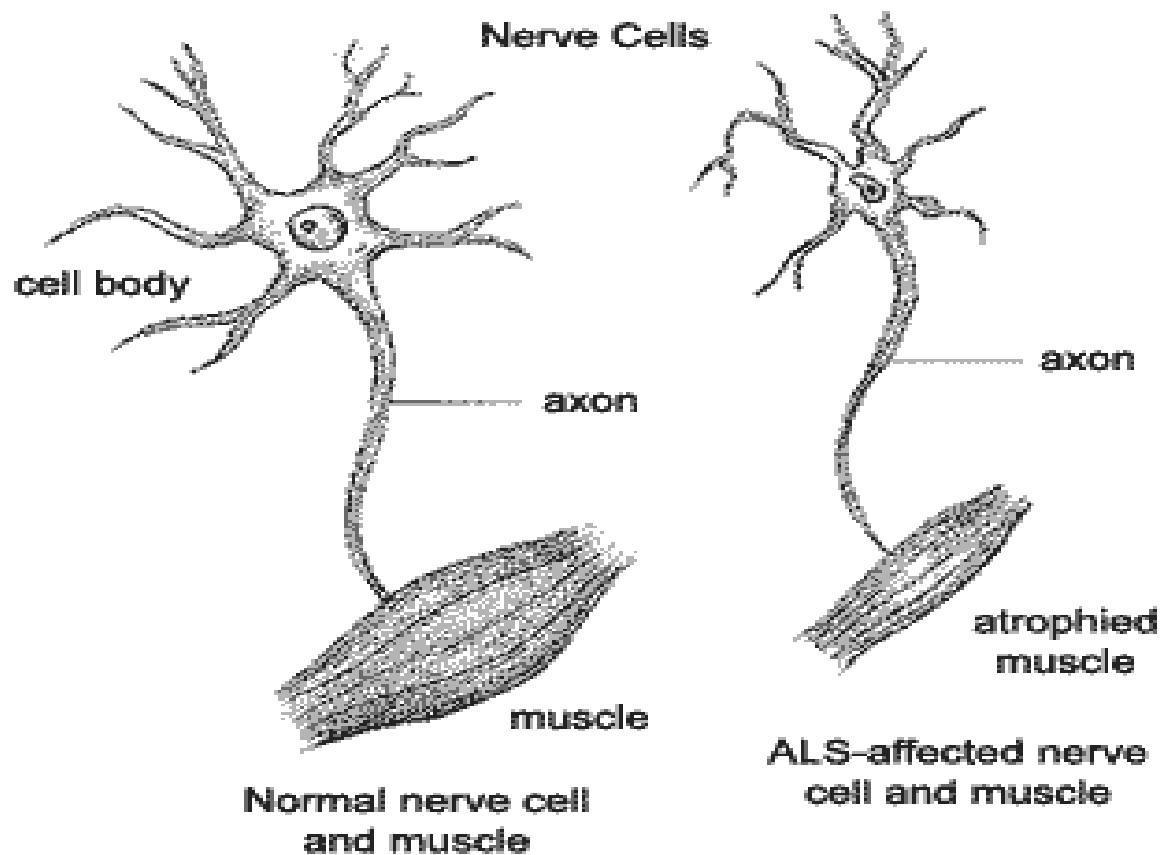
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# Understanding ALS



You, a family member, or a friend has been told that Amyotrophic Lateral Sclerosis (ALS) is now part of your life. ALS is unique in that it is not defined by stages and it affects each person differently. How ALS will affect your body and your life is individualized - including the rate of progression, the muscle groups affected, and the length of the illness. As you read on, please remember that the probability of these changes exist however, it will run a variable course. Although life expectancy averages about two to five years from the time of diagnosis, many people live with a high quality of life for five or more years. More than half of all patients live more than three years after their diagnosis. 20 percent of patients survive more than ten years and 10 percent will live 20 years. In some cases, ALS has stopped progressing and in a small number of cases symptoms have reversed. As your journey progresses, remember support is available through The ALS Association to help you remain in control of your healthcare decisions and communicate throughout your course of ALS. This assistance cares for you and your family as those around you will be journeying parallel with you.

Amyotrophic Lateral Sclerosis (also referred to as ALS, Lou Gehrig's disease, Motor Neuron Disease or MND) is a neurological disease, characterized by progressive muscle weakness and atrophy (wasting away or loss of muscle). ALS was first identified in 1874 by a French neurologist, Jean Martin Charcot. It is classified as a rare disorder/orphan disease by the World Health Organization, since 30,000 people in the United States are living with this diagnosis at any given time. The incidence rate of ALS is about equal to Multiple Sclerosis with approximately 5,600 people being diagnosed each year. However, worldwide over 500,000 patients including all genders, culture and ages live with the disease at any given time.

It was nicknamed Lou Gehrig's disease as Lou Gehrig, a baseball player for the New York Yankees, was the first person with notoriety diagnosed in 1939. There have been many other famous people who have had ALS, however since the illness is often fatal in a short timeframe, a consistent spokesperson has not been identified with ALS. The disease has cut short the lives of other such notable and courageous individuals as Hall of Fame pitcher Jim "Catfish" Hunter, Senator Jacob Javits, actors Michael Zaslow and David Niven, creator of Sesame Street Jon Stone, television producer Scott Brazil, boxing champion Ezzard Charles, NBA Hall of Fame basketball player George Yardley, pro football player Glenn Montgomery, golfer Jeff Julian, golf caddie Bruce Edwards, British soccer player Jimmy Johnstone, musician Lead Belly (Huddie Ledbetter), photographer Eddie Adams, entertainer Dennis Day, jazz musician Charles Mingus, musician Mike Porcaro of Toto, composer Dimitri Shostakovich and U.S. Army General Maxwell Taylor and many other courageous men and women who succumb to ALS every day.

ALS is not just Lou's Disease, it is everyone's. It appears unpredictably and sporadically throughout the world with no boundaries in gender, race, or socio-economic groups. ALS is considered a mid-life disease with the most common age of onset in the mid-50s, however it can develop as young as a teenager and well into the 80s.

ALS is a motor neuron disease. It affects the nerve cells that control muscles we can move voluntarily. For some undiscovered reason, nerve cells in the brain and spinal cord, known as motor neurons, gradually are damaged and die. The result is that the muscles do not receive the necessary messages, and, therefore, do not function properly.

The name Amyotrophic Lateral Sclerosis accurately describes the disease state:

- ◆ “**A-myotrophic**” comes from the Greek language and literally means "without muscle nourishment" -- the "starved" appearance of muscles weakened from disease.
- ◆ “**Lateral**” identifies the location of the affected nerve fibers that pass through each side of the spinal cord.
- ◆ “**Sclerosis**”, also from a Greek word, means "hardening" -- an apt description of the scar tissue along the sides of the spinal cord caused by the death of upper-motor neurons.

Amyotrophic Lateral Sclerosis means muscular weakness and degeneration of nerves. Motor neurons are among the largest of all nerve cells. They reach from the brain to the spinal cord and from the spinal cord to muscles throughout the body. When motor neurons die, as they do in ALS, the ability of the brain to start and control muscle movement dies with them.

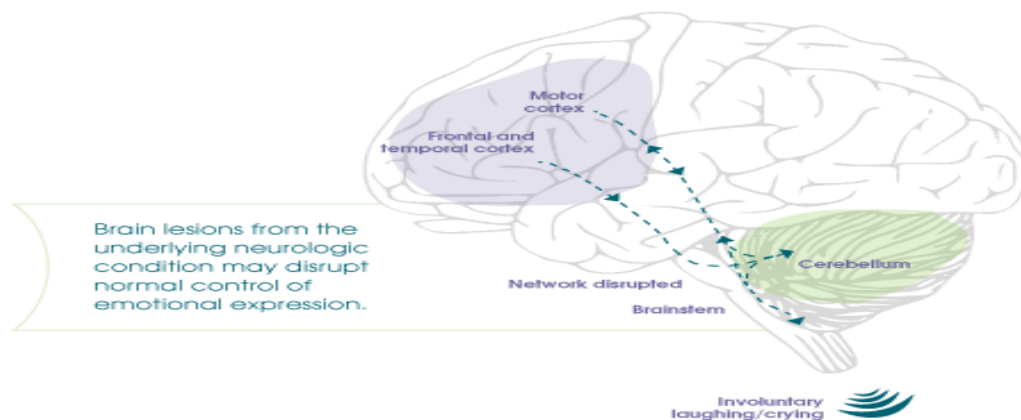
Consider what happens when a healthy person undertakes an action as simple as picking up a glass of water. First, the brain sends out electrical and chemical "messages" to nerves directed to instruct the hand muscles. Second, the motor neurons pick up these messages and provide a passageway for them to travel along to the hand. Third, after the motor neuron has transported the signals to the end of the passageway, it releases the signals to the hand muscles. Lastly, the muscles then recognize and respond to the command to grip the glass.

In the ALS patient, this sequence of events is eventually disrupted because the individual motor neuron is dead; it cannot produce and transport the vital signals to the muscle. Electrical and chemical messages, originating in the brain, never reach the muscles to activate them. Muscles then weaken from lack of stimulation and use.

ALS begins in one region of the nervous system that causes the upper and lower motor neurons to die in that area; then the muscles they control become weaker

and smaller. The strength of any voluntary muscle group can be affected in ALS, including those that control facial expressions, chewing, swallowing, speaking, the neck, arms, trunk, legs, and breathing. Bulbar Onset ALS is defined when the muscles controlled by the bulbar region located in the brain stem at the base of the skull begin showing symptoms first. Those muscles support chewing, speaking, and swallowing and aid with breathing. Limb onset ALS is when initial symptoms present in arms or legs. ALS can start in any muscle group and then move to any other, there is no clear pattern of the spread of weakness to other muscle groups.

ALS may also affect emotions, resulting in periods of inappropriate or exaggerated emotional responses. Traditionally, this pseudo bulbar affect is displayed through laughing and crying. It happens when certain neurological diseases or injuries damage the area of the brain responsible for controlling what we typically consider to be the normal expression of emotion. The damage can affect brain signaling, short circuiting the normal system and causing episodes of involuntary crying or laughing. The outpouring of emotion may be out of context with the person's actual feelings. As many as 15-45 percent of patients, may experience pseudo bulbar affect.



Muscles such as those that move the eyes and the external sphincters that hold stool in the bowel and urine in the bladder are very rarely involved in ALS. Other

areas not affected are internal organs (such as the heart, liver, and kidneys), sexual functions, and the five senses (sight, hearing, smell, taste, and touch/pressure).

Traditionally thought that the the mind and mental abilities are rarely affected by ALS, researchers have begun to recognize an important connection between fronto temporal degeneration (FTD) and ALS. FTD is a syndrome of progressive changes in behavior and language due to loss of function of neurons in the frontal and temporal lobes of the brain. In many cases, FTD symptoms are noted prior to the diagnosis of ALS. Most patients will remain physically strong and relatively agile until late in the illness. 10-15 percent of FTD patients, develop damage in the nerves cells that control voluntary movement or motor neurons. When this occurs, the syndrome is called FTD/MND or FTD with ALS.

FTD is a form of dementia, or altered mental status, which is distinct from the cognitive and memory problems evident in other dementia diseases. FTD involves a change in personality that can produce obsessive behavior, or the loss of the ability to plan or make decisions. Fronto temporal refers to the forward part of the brain that sits above the eyes and behind the temples. Lowering of the function of this region can lead to impulsive and emotional behavior. Patients with this diagnosis usually experience a rapid decline in both physical and cognitive abilities. Approximately 30% of ALS patients will show signs of frontal lobe decline, which affects organizational function and behavioral comportment.

The most common form of ALS in the United States is sporadic ALS with 90-95 percent of all cases having no familial link.. Subcategories of ALS exist when symptoms are present in one muscle group for an extended period of time. Because of the progressive nature of ALS, these sub-categories; possible, probable, upper/lower motor neuron disease, spinal or progressive muscular atrophy (SMA), progressive bulbar palsy (PSP), or primary lateral sclerosis (PLS) traditionally overtime loss of function expands to other muscle groups and are



often re-diagnosed as ALS. However, a timeline cannot be estimated for this reclassification as ALS is individualized from person to person and some cases do not evolve into ALS.

Genetics accounts for 5-10 percent of ALS cases. In 1993, a team of scientists led by ALS Association funded researchers discovered the first gene that, when defective, causes most cases of familial ALS (FALS). This defective gene, which appears to be the cause of about 20 percent of all familial ALS, was linked to chromosome 21 and is known as SOD1. This results from mutations in the gene that encodes the enzyme copper – zinc superoxide dismutase. More recent research advances has shown that there a dozen common genes with mutations currently known to be associated with FALS including; SOD1, TDP-43, TARDBP, FUS, ALS6, UBQLN2, and C9ORF72. About one-third of all familial cases (and a small percentage of sporadic cases) result from a defect in a gene known as “chromosome 9 open reading frame 72,” or C9orf72. The function of this gene is still unknown. ALS6 is responsible for 5 percent of FALS cases.

There are several inheritance patterns, but the most common inheritance pattern for FALS is called autosomal dominant. Autosomal means that it is equally likely that a female or male would inherit the gene mutation for FALS because the gene is located on an autosome – a chromosome that both males and females share in common. Dominant refers to the fact that a person only needs one gene to have a mutation in a gene for FALS to have an increased risk for ALS. Someone who has FALS would have one copy of the gene with a mutation and one copy of the gene without a mutation. Therefore, a child born to someone who has FALS has a 50% chance to inherit the FALS gene mutation and conversely, a 50% chance to not inherit the FALS gene mutation. This 1 in 2, or 50% chance, comes from the fact that parents randomly pass on only one member of their gene pair, so that either the gene with the mutation will be passed on or the gene without the mutation will be passed on. Even though parents often feel responsible for their children's

health, they have no control over which gene they pass on, just as their parent had no control which gene they passed onto their child. It is also important to remember that inheriting the gene for FALS in no way guarantees that a person will develop symptoms of ALS. Also, if a child does not inherit the gene mutation for ALS, they cannot pass it onto their children.

Genetic counseling is available for FALS patients and families. The genetic test is typically a blood draw and is sent to a special lab for sequencing and mapping. Options for prenatal genetic counseling also exist. These tests are not typically covered under insurance. This is a highly controversial and personal topic of discussion that requires at least a year of genetic counseling because a positive genetic test at this time would not change medical intervention or prevention risk factors. It could carry serious psychological ramifications since it is still impossible to detect at what age ALS MIGHT develop.

In 2000, Researchers around the world united to create an ALS Consortium. This consortium created a strong network of information sharing between three main research groups. One group focuses on detecting the cause of ALS, the second group researches ways to slow down the progression, and group three's focal point is identifying a cure for ALS. Since this time, research information has been shared and has advanced our understanding of ALS.

In 2008, Congress enacted the [ALS Registry Act](#), to create and maintain a registry of persons with ALS. The registry launched in October 2010 and is maintained by the Centers for Disease Control, seeks to help researchers estimate how many people have the disease, understand more about who gets the disease and potentially find a cure. To learn more, or register, [www.cdc.gov/als](http://www.cdc.gov/als)

The ALS Association currently sponsors research projects around the world and is the global leader in research efforts. The ALS Association's TREAT ALS [Translational Research Advancing Therapy for ALS] combines efficient new drug

discovery with priorities set for existing drug candidates, to accelerate clinical testing of compounds with promise for the disease. “Now tangible progress will be turned towards patients to produce treatment success,” according to Lucie Bruijn, Ph.D., The ALS Association’s science director and vice president. “Translational research and clinical trials will find the drugs which will prevent, halt, or significantly slow down disease progression.” “Our ultimate goal is to capitalize on scientific and technological progress to accelerate drug discovery and realize effective new therapy,” said Lucie Bruijn, Ph.D., ALS Association science director and vice president. “We understand far more about the biological basis of the disease and this knowledge has enabled design of laboratory models of ALS that have yielded innovative ideas and novel treatment strategies. Already partnering with many organizations around the world including The National Institutes of Health (NIH), the largest single investor in research globally, The ALS Association brings together an expert team of scientific and business advisors to steer this initiative. “As part of this initiative we will support the development of novel compounds for large scale, U.S. Food and Drug Administration approved clinical trials and engage in small pilot trials of existing FDA-approved drugs,” said Bruijn.

Speculation about the causes of ALS has prompted consideration that ALS may not have a unitary cause - it may represent an end-state reached through more than one pathway. If this is correct ALS treatment may require a multi-model strategy.

We recognize that genetics play a major role in ALS as well as how genes interact with the environment. Another area that has become of particular interest in this challenging disease is the field of biomarkers, which are signatures that differentiate between people who have the disease and those who don’t. This is important for two reasons – one is the possibility for earlier diagnosis. A major challenge for treatment has been that by the time a person’s ALS diagnosis has been confirmed about 50 percent of the motor neurons have already died. Earlier diagnosis would allow treatment to begin earlier, and hopefully, prevent cells from

dying. The other reason biomarker research is important is for what it could offer clinical trials. Biomarkers can track whether a drug is, in fact, making a difference.

Researchers are looking for aspects of lifestyle that can interact with genes or cause or contribute to ALS. Exposure to toxins or the influence of intense exertion, are ideas researchers consider as possible reasons for the finding that some veterans and some athletes have increased incidence of ALS.

An ALS Registry for Veterans was established in 2005. A recent study found that the relative risk of dying from ALS for veterans was 1.5 times greater than those who did not serve. In addition the rate of ALS in young Gulf War veterans was more than two times greater than expected in the general population. If you are a veteran, please refer to the Veterans with ALS Manual ([put in link](#))

As of yet, no conclusive proof exists for any toxin as a causative factor in ALS. Areas that scientists have researched include heavy metals, solvents, radiation, and electromagnetic fields. One of the first clues that ALS might involve an environmental factor was obtained on the island of Guam in the Pacific where an unusually high proportion of people over the past century have developed symptoms similar to ALS as they age. No proof has yet nailed down any dietary deficiencies or excess as a cause of ALS.

Many investigators have looked at heavy metal exposure, particularly lead, and including mercury and manganese, as risk factors for ALS. A positive association between past exposure to heavy metals and risk of ALS has not consistently appeared across studies, including the regions of Jefferson County in Missouri and in Jackson County, in Illinois. Environmental influence is another reoccurring theory on ALS. This speculates that a virus is responsible for the condition. A concrete link to viral infections has never been documented for ALS or other disorders of the nervous system.

Laboratory models of ALS helps researchers understand the basic process of the disease. The mainstay has been a mouse that bears the mutated human gene associated with familial ALS. A newer rodent model, an ALS rat, also is engineered to express human mutant SOD1. The rat is large and surgery is easier for applications such as stem cell transplants. The worm, fish, and the fly models of ALS will be especially valuable, even as cells growing in a dish. Cell based tests that reflect the disease process in ALS can rapidly report on the potential of new molecules to serve as therapies.

The discovery that human embryonic stem cells can be isolated and propagated in the lab with the potential of developing into all tissues of the body is a major medical breakthrough, but it has raised ethical concerns. If there were a way to stimulate resident stem cells to replace dying cells, the limitations of transplantation could be overcome, as well as the ethical issues. For ALS, it is becoming evident that it is not only the motor neuron that is at risk in the disease but neighboring cells as well. Attempts to replace these cells are ongoing and may be more feasible than motor neuron replacement. In the immediate future, stem cells may be vehicles that can be sent to the damaged area and provide missing factors to help remaining cells survive.

Significant progress has been made in the study of ALS. Although there is still no cure, recent clinical trials have shown that some drugs affect nerve cell activity and may increase the survival time for people with ALS. Newly developed animal models of the genetic form of this disease, so-called transgenic ALS mice, offer neurological researchers the ability to test therapies in mice. There is great hope that this and other neuro-scientific advances will lead to a cure in humans. You may find information on the National ALS Association website regarding stem cell research studies by visiting [www.alsa.org](http://www.alsa.org) and access the research tab. The National ALS Association has a current detailed listing of all ongoing research

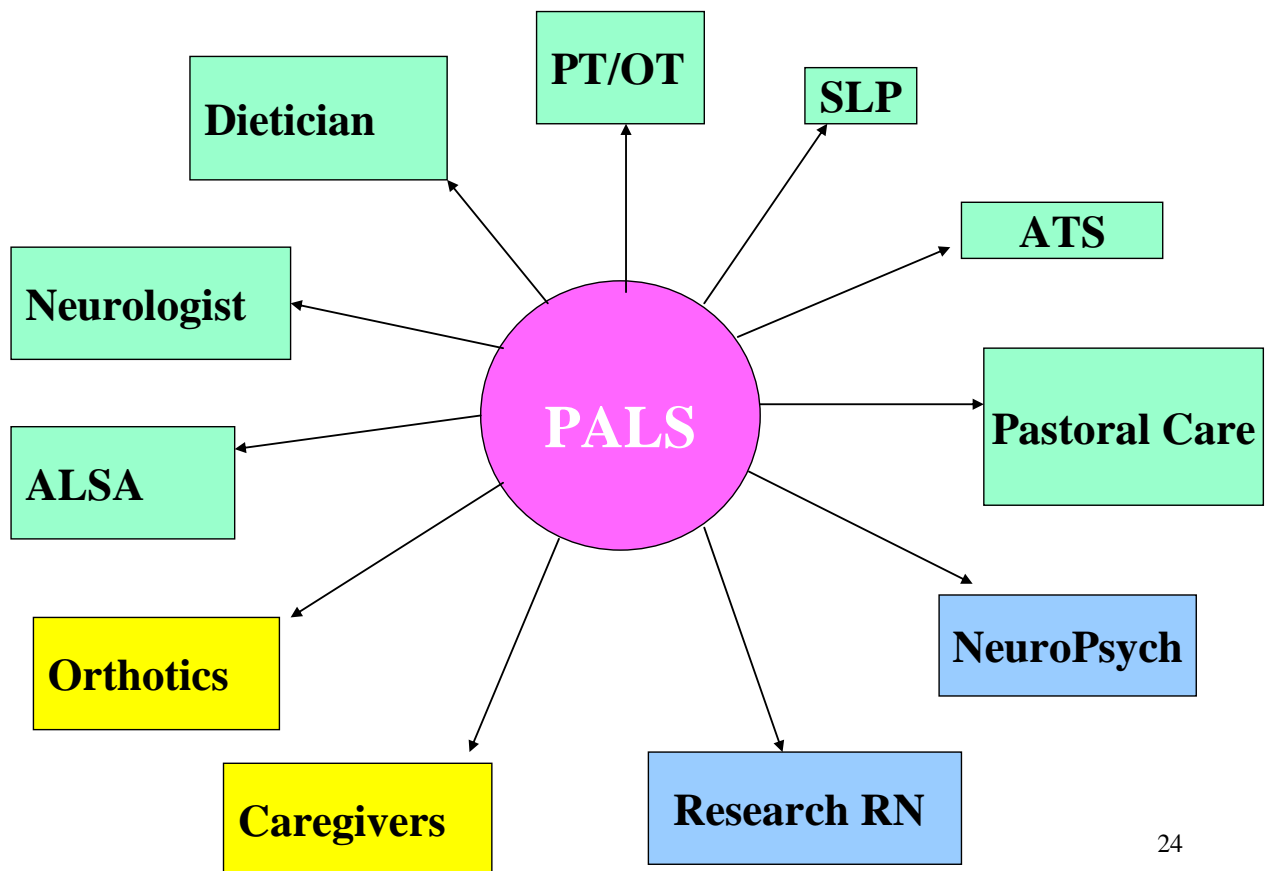
studies, listed on their website at [www.alsa.org](http://www.alsa.org). You may search this site by topic, location, or researcher. If you do not have internet access, call the St. Louis Regional Chapter office and an updated comprehensive listing can be sent to you.

## The Amyotrophic Lateral Sclerosis Association's Patient Bill of Rights for People Living with ALS

As a person living with ALS, you have the right to:

1. Receive comprehensive information about ALS, including treatment options and resources for your health care needs. This includes the right to communicate with your government representatives regarding policies and practices of the Food and Drug Administration (FDA), National Institutes of Health (NIH), Department of Health and Human Services (DHHS) and other agencies that impact ALS.
2. Participate in decisions about your health care including the right to accept, discontinue or refuse treatments and therapy.
3. Receive ALS specialty care in a timely manner.
4. Receive health care that is coordinated and individualized for you across the spectrum of home, hospice, hospital, nursing home, outpatient, and work-place and throughout all the phases of your illness.
5. Access health care benefit coverage and life insurance coverage without discrimination based on your ALS diagnosis or disability.
6. Obtain clear, timely information regarding your health plan including benefits, exclusions and appeal procedures.
7. Review your medical records and have the information in your records explained to you.
8. Prepare an advance directive to state your wishes regarding emergency and end-of-life treatment choices.
9. Receive care that is considerate and respects your dignity, your cultural, psycho-social and spiritual values and your privacy. You have this right no matter what choices you make about treatments and therapy, what your disabilities related to ALS might be or what your financial circumstances are.
10. Know that information about you and your medical condition are held in confidence.
11. Receive support to maintain or enhance the quality of your life and have your family involved in all aspects of your health care.

# Supportive Therapy & Symptom Management





ALS is a very difficult disease to diagnose. There are several diseases that have some of the same symptoms as ALS .

Symptoms experienced are:

- fatigue, loss of muscle strength and/or weakness in hands, leg, etc.
- tripping, foot-dragging; muscle twitching (fasciculation)
- cramping or stiffness in muscles and spasticity
- slurring of speech or difference in quality of voice
- trouble with chewing, swallowing, tongue, lips, etc.
- inability to take deep breaths or shortness of breath
- inability to perform some movement or task once easily done
- progression or worsening of symptoms
- cognitive or behavioral change
- emotional incontinence or overload

To date, a single test or procedure to diagnose ALS has not yet been developed. On the average, it takes approximately nine months to complete a battery of tests to rule out other neurological disorders that have the same symptoms as ALS.

Routine tests to establish a diagnosis of ALS include:

- **EMG** (electromyography) shows a muscle's electrical activity, and can show abnormal movement. This is an invasive test where needles are placed through the skin into the muscle.
- **NCV** (nerve conduction velocity) is the time it takes for electrical impulses to travel a length of nerve. The results of this test are usually normal for ALS, but the test can indicate other disorders.
- **Blood tests** are a necessity for a complete physical and may indicate other conditions.
- **Urine tests** involve a 24 hour collection to check for any metals in the system.
- **X-ray** include computerized axial topography (CAT), single photon emission computed topography (SPECT or PET), and magnetic resonance imaging (MRI).

They can show early symptoms that can be attributed to pressures on the spinal cord by tumors, cervical spine conditions, etc.

- **Spinal taps** involve a drawing of spinal fluid to be examined for elevations of proteins that are suggestive of motor neuron disease. This is another invasive test using a long needle inserted into the spinal canal at the lumbar area.
- **Muscle biopsies** are also invasive, with a small piece of muscle taken from an arm or a leg for microscopic examination. This is usually done with unusual symptoms or difficulties in diagnosis. After this test, a person may feel pain.
- **Pulmonary-function** tests check breathing capacity and provide a baseline for progression of respiratory involvement.
- **Barium Swallow** tests assess a person's ability to swallow and measures the proper consistency for safe eating and drinking.

Prior to diagnosis, it is not uncommon for ALS patients to have unnecessary spinal surgeries, carpal tunnel releases, or stroke therapy, all the while muscle weakness progresses. By the time a diagnosis is made, the patient and patient's family is frustrated with the medical community. However, it is important to continue pursuing treatment as symptom management can help to increase your quality of life and maximize your functional independence. The ALS Association recommends that every ALS diagnosis get a second opinion by an ALS expert, a neurologist who diagnoses and treats many ALS patients and has training in this medical specialty. These physicians are most commonly found in one of the 34 ALSA Certified ALS Centers of Excellence multidisciplinary ALS clinics or within one of the 53 recognized treatment centers around the US. For a comprehensive listing, please visit [www.alsa.org](http://www.alsa.org) or call the St. Louis Regional Chapter's office. A listing of all multidisciplinary clinics in IL, MO, Kansas, Indiana, Kentucky, and Arkansas is included at the back of this section.

The 2009 American Academy of Neurologists Practice Parameters Update, a document that establishes the treatment protocol for ALS patients, supported

earlier documentation that a multidisciplinary clinic referral should be considered for managing patients with ALS to optimize health care delivery, prolong survival and enhance quality of life. In 2013, Forbes Norris conducted a similar study to that of the Professor Leonard van den Berg of the Netherlands, with similar results – that the quality of life is for the person with ALS/MND does improve by being treated at a multidisciplinary clinic or center. The multidisciplinary approach pulls a team of allied health professionals to offer comprehensive treatment in a compassionate setting. The team treats not only the ALS illness, but the person who has ALS, and the family supporting that person. In a 2013, the ALS Worldwide Organization reported that these multidisciplinary centers and clinics and concluded that these clinics provide excellent care to ALS/MND patients, from the patient's perspective.

Although it is impossible to erase the weakness and progressive muscle paralysis that accompany ALS, team members strive to maintain muscle function and ability to perform the daily activities of life and those activities that help a person live longer and stronger while journeying with ALS. The primary treatment remains the management of ALS symptoms. Patients need to take an active role in the design of their treatment regimen.

In general, the person with ALS needs the following:

- ◆ Techniques for managing daily symptoms
- ◆ Ongoing teaching and counseling
- ◆ Straightforward information about the disease process
- ◆ Access to equipment and services
- ◆ Timely evaluations of the progression of the disease

# **T**HE MULTIDISCIPLINARY ALS HEALTH CARE TEAM

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**NEUROLOGIST** - The physician works with the members of the healthcare team to make sure you receive the specialized care you and your family needs. The neurologist will also work with your primary care physician to keep him/her informed of the current treatment and care.

**NEUROMUSCULAR RESEARCH NURSE** - At all multidisciplinary clinics, ongoing research projects are available for patients to enroll. It is an important option to consider as it may make a difference in the course of your illness and those that will be diagnosed after you.

**DIETICIAN, NUTRITIONIST** -- A well-balanced diet is an important consideration for ALS patients. As the disease progresses, difficulties in maintaining good nutrition will be encountered. The dietitian's primary considerations are to keep the individual's quality of life as high as possible by maintaining safe and adequate nutrition and hydration. This will prevent life-threatening nutritional deficits from developing.

**OCCUPATIONAL THERAPISTS (OT) and PHYSICAL THERAPISTS (PT)** play an important role in the overall wellbeing for the ALS patient. OTs and PTs develop management strategies that enable patients to continue to carry out work and leisure activities in a safe and efficient manner. The occupational therapist is generally more concerned with fine-motor control and the application of movement to function. The physical therapist is generally more concerned with gross-motor function and anatomical impairments associated with abnormal movement. The PT and OT will work together with you in your home to provide

gait and equipment training, ROM exercises, energy conservation, transfer training, and assess your home for safety. Finally, occupational and physical therapy services can be provided in the home.

**PASTORAL/SPIRITUAL CARE** -- Psychological manifestations such as denial, anger, withdrawal, and depression are common and normal reactions of both the patients with ALS and family members. The patient's psychosocial-spiritual status is of great importance and profoundly affects the patient's experience. It is important to address end of life concerns and issues early in the disease process when energy is high and the voice is strong. In home services are available to supplement this support.

**RESPIRATORY THERAPIST AND PULMONOLOGIST** -- The respiratory therapist generally devises strategies to optimize remaining muscle function and reduce discomfort. They offer information on body positioning, energy conservation, relaxation, and compensatory techniques to improve breath support for nutrition and for speech. They also assist with monitoring the vital forced capacity and pulmonary function scores. These are key pieces of medical information that will assess the disease course and diaphragmatic involvement that will help you make a choice regarding ventilator support. Non-invasive breathing support is also available when patients experience breathing or sleep difficulties. This information is important for your medical team regarding the decision to refer to Hospice.

**ASSISTIVE TECHNOLOGY SPECIALIST** - Many devices, pieces of medical equipment, and special switches can enhance a person's functional independence and maximize their overall quality of life. It is important to remember that using such devices will not speed up the progression of ALS. Even though it may seem that using an electric wheelchair is a defeat, it is important to focus on the freedom

it will give back to you. Enclosed at the end of this section is a CD-ROM of such devices.

**SOCIAL WORKER** -- The social worker may promote an understanding of and adjustment to the disease process, share information about available community resources, provide information on legal matters such as advanced directives and living wills, assist in making long-range plans, provide counseling and guidance, assist caregivers, and give emotional support.

**SPEECH-LANGUAGE PATHOLOGIST** -- The speech-language pathologist provides advice on compensatory techniques and augmentative communication strategies and devices that allow the individuals to continue to communicate. They also provide training in safe eating, drinking, and swallowing techniques. In home services are also available to offset this support.

**ORTHOTIST** - The orthotist will help you maintain comfort and mobility through the use of special braces. The most common braces include leg, neck, ankle and foot. Most braces are individually fit to each patient to maximize comfort and function. Braces are typically needed for your limbs.

**ALSA CARE SERVICES CASE MANAGER**- functions as the eyes and ears of the team in-between clinic visits. The Care Services Specialist will work with you and our family in your home to make sure all resources, both ALSA, ALS specific, and other community resources are utilized. The Care Services Specialist will also have information on all areas of ALS and how it affects a person's life.

Once a diagnosis has been confirmed with a second opinion, it is important to reflect on your decision regarding end of life choices. At some point, most ALS patients must consider the placement of a feeding tube and invasive breathing support. By completing a living will and appointing someone that understands

your choice as a proxy or Durable Power of Attorney for Healthcare, you ensure that you are in control of your healthcare decisions. Although these are difficult decisions to make, it is important to let wishes be known. It will empower your family's decision making ability and confidence that they are following your wishes at a very difficult time for all. Samples of these documents are included at the end of this section.

Nontraditional treatments differ from experimental or unconventional and include; nutrition, homeopathy, acupuncture, healing massage, chiropractic, visualization techniques, and faith healing may play important roles in ALS care. Discuss all treatments (conventional and unconventional) with your ALS doctors to make sure that no harm will be done to you and to determine what advantages may be obtained. Other types of alternative treatments and drugs may be unorthodox and do not have FDA approval. Again, be cautious before trying a new drug or treatment not prescribed by your healthcare providers.

In 1992, the FDA approved Rilutek (Riluzole) as a treatment option for ALS patients. Laboratory studies suggest that Rilutek is "neuroprotective" which means it protects nerve cells from damage. Many researchers believe that Rilutek protects the nerve cells by what they describe as "glutamate inhibition." The human body naturally produces a substance called glutamate. Glutamate carries signals to the motor neurons. This is one link in the chain of how the brain tells the muscles what to do. One of several theories researchers have is that in people with ALS, parts of the nervous system are overexposed to glutamate. A buildup of glutamate has been seen early in the progression of this disease. Too much glutamate "burns out" the motor neurons so that they can carry the brain's messages to the muscles to tell them what to do. Although researchers don't fully understand how Rilutek works, one explanation is that Rilutek may protect nerve cells from overexposure to glutamate. Unfortunately, not all ALS patients have an excess amount of glutamate and medical technology has not produced a

test to detect this. The clinical trials research proved the Rilutek slowed down the progression in ALS patients taking the drug vs. those taking the placebo by 3-6 months. It is not known where in the course of ALS that falls but it did indicate that taking Rilutek as close to the time of diagnosis the results tended to be stronger. On the average, the cost of Rilutek ranges from \$200 – 500 per month for the generic and up to \$2000 per month for the namebrand. If you are joining Medicare Part D, it is covered under most plans.

Some ALS patients at times seek alternative or unorthodox forms of treatment, those outside of the traditional medical settings. Sometimes these non-traditional therapies will show a temporary improvement based on the placebo effect or an initial system response falls back to the pretreatment state eventually. The tendency is for patients to say why not try it, I have nothing to lose. This can be a dangerous approach for several reasons: Patients are removed from the mainstream of what medicine does have to offer by way of symptomatic treatment and clinical trials, patients are lead to physical and financial sacrifice and eventual failure leads to an inevitable psychological let down.

Alternative treatments and drugs may be unorthodox and do not have FDA approval. Again, be cautious before trying a new drug or treatment not prescribed by your healthcare providers. ALS patients have spent a great deal of money and hope into these treatments with little concrete and lasting effects. Some examples are listed below:

- Mega doses of vitamins, minerals, and supplements.
- Stem cell treatments and transplants in China and Mexico
- Snake venom
- IV treatment that bridges the gap between western medicine and alternative medicine. The concept is to stop the damage at the cellular level and rebuild the myelin sheath around the nerves by giving supplements and, “other stuff.”



- High doses of antibiotics.

Although the field of stem cell research offers insight and hope into ALS, it is not considered a treatment for ALS patients at this time. Stem cells are cells that have the ability to divide for indefinite periods in culture and give rise to multiple specialized cell types. Embryonic stem (ES) cells can develop into any of the tissues that form the body and are considered to be pluripotent. Earlier studies focused on mouse ES cells, however recently scientists have shown that they are able to isolate and propagate human embryonic stem cells in culture. This breakthrough has fueled much excitement with the hope that the use of human stem cells may reduce the chances of tissue rejection after transplantation. However, as these cells are derived from human embryos, several organizations have opposed these studies. Pluripotent stem cells undergo further specialization into multipotent stem cells that give rise to cells with a particular function. For example, multipotent stem cells in the brain give rise to different neuronal cell types and glia. While stem cells are important in early human development, they persist into adulthood. Their function in adulthood is less clear. The presence of bone marrow stem cells in adults has been known for a long time. These stem cells give rise to all cells of the blood system. More recently, stem cells have been discovered in the adult brain and spinal cord.

There are specific challenges that face Stem Cell Therapy in ALS. The discovery that human embryonic stem cells can be isolated and propagated in culture with the potential of developing into all tissues of the body is a major breakthrough for the field. However, it has raised many ethical questions. The NIH currently funds research using embryonic stem cells; however, funding is restricted to cell lines that are approved under the NIH guidelines. One of the major concerns among investigators is the limitation of these cell lines and the need to generate new more appropriate cell lines. There are several efforts underway within the U.S. and abroad to develop new lines outside NIH funding. An intriguing

discovery is that bone marrow cells (which are able to develop into all the cells of the blood system) transplanted into mice can migrate into the brain and develop into cells that appear to be neurons. These studies remain controversial, however, as scientists dispute the capacity of these bone marrow cells to efficiently generate neuronal like cells. They may be effective in providing the necessary trophic factors for motor neuron survival. Studies continue in this field and, if successful, this would be the most convenient source for stem cell therapy.

The mechanism of motor neuron death in ALS remains unclear. It is not certain that transplanted stem cells are resistant to the same insult(s) causing motor neurons to die and stem cells may need to be modified to protect against the toxic environment. Embryonic stem cells in culture can be genetically modified. An attractive use of stem cells is as a vehicle to deliver genes and other factors to dying motor neurons. More research is needed in this area.

Despite encouraging data that transplanted fetal cells can survive over long periods of time in the damaged area, few studies have shown functional recovery of neurons (neurons making appropriate contact with their target). In addition, unlike Parkinson's disease where functional improvement is less dependent on appropriate neuronal connections, motor neurons have a huge challenge to form connections with their target (muscle) over a very long distance.

The presence of neural stem cells in the adult brain and spinal cord may provide an alternative to transplantation eliminating the issues of tissue rejection. If there was a way to stimulate resident stem cells to replace dying cells, the limitations of transplantation could be overcome. Small biotech companies are pursuing this direction in the hope of finding therapeutic compounds that will do this. Further research into molecules and genes that govern cell division, migration and specialization is needed, ultimately leading to new drug targets and therapies for ALS.

The following pages address some of the main symptom management treatment protocols associated with ALS. Whether your ALS is eight months, eight years, or eighteen years, these concerns are typically affected to some degree.

As you continue to read, please remember that The ALS Association is available to assist you and your family through these issues.

## **Mobility Issues with ALS**

Sometime throughout the course of ALS, most patients will have difficulty with ambulation. Because ALS is so individualized from person to person, this progression may happen more rapidly (within six months) or take a slower course (over several years). Loss of balance due to foot-drop, muscle atrophy, and spasticity can make walking extremely difficult and dangerous. Safety must become a primary concern. Living with ALS is challenging enough without the added burden and pain of injury. Remember, ALSA chapters loan equipment.

There are several mobility aids that can help ALS patients maintain independence, conserve energy, and avoid consequences of a serious fall and related injury. An occupational therapist and or a mobility specialist can help you determine the right time frames and options to choose. Medicare and most insurance carriers cover these expenses; however it is important to review your certificate of coverage. Some will only cover one transportation or mobility aid.

**The Ankle Foot Orthosis (AFO):** One common mobility symptom resulting from ALS is the inability to hold the toe of one or both feet up while walking. This is commonly referred to as foot-drop and results in the patient having to lift the foot more than normal while walking to avoid tripping. Correcting foot-drop with a lightweight ankle-foot orthosis can be helpful to minimize falls and maintain endurance. Although these are available off the counter, it is important to get measured properly for safety and independence concerns.

**Canes:** The standard cane has a single tip while the quad cane has a rectangular four-tip base for improved stability - which style works best for patients will vary depending on conditions and can only be adequately determined by having the patient try each style.

**Walkers:** come in many styles and designs with features such as: three or four wheels, hand brakes, baskets for carrying items, and fold down seats.

**Manual Wheelchairs:** can be custom-fitted or are available with standard options. Can be self propelled or pushed by another individual.

**Scooters:** Because of its costly price and it is typically useful for only a short period of time, it is advisable for ALS patients to skip a scooter and begin using a power wheelchair.

**Power Wheelchairs:** come in a large variety of designs/options. A suitable wheelchair for an ALS patient will typically cost in excess of \$15,000. Insurance companies often deny expensive equipment and will commonly fight to purchase less expensive/less adequate equipment. You will likely be required to justify the medical necessity for the wheelchair and options and features you need. For this reason, it is important to have your wheelchair prescription prepared by a physical therapist/mobility specialist who understands ALS and can specify/justify your requirements in detail. Optimally, this evaluation should take place in your home, to view doorways and accessibility. It takes four-eight weeks to obtain a power chair so plan ahead and think about accessibility of your home. **See the brochure at the end of this section for recommended options and providers.**

**Pain Management and Prevention Techniques:** Many people believe that ALS patients experience no physical pain as a result of their disease but many PALS experience varying degrees of pain. The origin of the pain comes from several sources: muscle cramping, spastic muscles, joint pain due to immobility, pain from pressure points, and poor body positioning. It is important to identify the source of the pain in order to treat it effectively. It is also possible to prevent some of the pain associated with ALS with proper medical and therapeutic management. Pain pills can be used in conjunction with some of the

management techniques described below as needed but will not alleviate the source of the pain. **DISCUSS ALL PAIN WITH YOUR PHYSICIAN!**

**Muscle Cramping:** This is quite common in early and middle stages of the disease and can affect any muscles. Muscle cramps can be very painful and are caused by an over contraction of the muscle, probably due to misfiring nerve impulses. They tend to be triggered by overusing the muscles, cold temperatures, and decreased circulation. Once the cramping response is triggered it can last for a while. To prevent cramping try to pace your activities so as not to overexert your muscles taking frequent rest breaks, avoid cold temperatures, and participate regularly in a light exercise program to improve circulation. Quinine is a drug, which is effective in reducing muscle cramping. Muscle relaxants are used but you need to work with your doctor on the appropriate dose because too much can cause muscle weakness.

**Hypertonic or Spastic Muscles:** Spasticity is not a symptom for all PALS and is a function of upper motor neuron involvement. Spasticity is defined as increased tone or contractions of muscles causing stiff, awkward, and jerky movements. If a PALS with spasticity tries to move, is touched, or is positioned in a certain way an exaggerated motor response often occurs. This can take place in the limbs and/or the trunk causing the affected areas to stiffen which can be painful if severe. Proper positioning in bed and in a wheelchair can inhibit spastic responses as can the prescription drug Baclofen which is very effective in decreasing spasticity. A PT or OT can offer advice for proper positioning techniques.

**Joint Pain and Stiffness:** As strength is lost, it eventually is not possible to move each joint through its complete range of motion on a daily basis. Over time, if the joints remain immobile the muscles, tendons, and ligaments will shorten causing joint pain and restriction of movement. This type of pain can be easily prevented by adhering to daily range of motion exercises which can be taught to you and

your caregiver by a PT or OT. ROM exercises for all joints of the limbs as well as the neck are very important. Many PALS also take daily doses of prescription strength anti-inflammatory to help relieve joint pain effectively. These can be hard on the stomach so you may also need to take an acid blocker as well.

The shoulder joint can be especially problematic. As shoulder muscles weaken, the shoulder joint can sublux (mildly dislocate) in addition to becoming restricted in motion. The connective tissue around the joint can also become inflamed and a continual cycle of pain can develop. In order to prevent shoulder subluxation and inflammation it is essential that weakened shoulders are supported and properly positioned. A PT/OT can show you how to achieve this along with daily shoulder range of motion exercises. If you already have a painful shoulder, it can be treated effectively with help from a PT/OT using deep heat or ice and mobilization techniques in addition to anti-inflammatory/pain medications.

**Painful Pressure Points:** If a PALS remains in one position for hours on end while in bed or sitting in the wheelchair pain can develop from continual pressure on bony areas like the hips, heels, and tailbone. Regular repositioning, an alternating pressure pad, and special foot positioners used at night can prevent pain from pressure points in bed. Also special back and seat cushions are effective in preventing painful pressure areas while seated in a wheelchair along with the ability to recline and tilt the wheelchair in order to achieve pressure relief.

**Poor Positioning:** As muscles weaken special positioning devices may be needed to maintain normal alignment of the body and prevent pain. Hand splints, neck braces, lumbar supports, etc. may be appropriate to accomplish this. Also special positioning devices on your wheelchair like lateral supports and neck rests are also important. A PT and OT can evaluate your positioning needs and make appropriate recommendations.

## **FEEDING TUBES**

A feeding tube is a small, flexible tube, about the diameter of a pencil, used to allow liquid nourishment to enter the stomach directly, bypassing the mouth, throat and esophagus. The feeding tube is often called a PEG tube, which stands for percutaneous (through the skin) endoscopic (into the GI tract) gastrostomy (hole in the stomach) tube. It's also simply called a gastrostomy tube, or g tube.

Many people who have ALS will develop dysphagia (difficulty swallowing) at some point. Swallowing can go from adequate to significantly impaired in months. This can lead to malnutrition and weight loss from inadequate protein and calorie intake. When this occurs, the body uses muscle as a source of energy and protein. This accelerates the progression of weakness. One of the risks of dysphagia is that food or liquid taken through the mouth may go down the trachea (windpipe), rather than down the esophagus to the stomach. This is called aspiration. Aspiration causes a high risk of infection in the lungs (pneumonia). Thin liquids and foods that crumble are especially likely to be aspirated. A feeding, or PEG, tube can restore adequate protein and calorie intake

When dysphagia symptoms begin, consider the placement of a PEG sooner rather than later because of nutritional deficiencies and the risk of aspiration. Another reason for placing the PEG early is that progression of respiratory weakness often parallels with dysphagia. When respiratory function is better, the procedure can be done with less difficulty and less risk of respiratory problems immediately following tube placement.



The gastrostomy tube creates an alternate route of entry into the stomach but doesn't affect oral intake. In the early stages of dysphagia, it can be used to supplement oral intake of food and fluids. Later on, when it's dangerous or impossible to consume any food or drink orally, it can be used as the sole method of nutrition. Many readily available liquid preparations designed to provide a complete and balanced diet can be used as the sole source of nutrition. (Ensure, Nestle or Jevity are a few brand names)

When a person has trouble swallowing, he may be unable to swallow pills. Many medications can be crushed, dissolved and given through the PEG. Some capsules can be opened and sprinkled into water or liquid food. Medication taken this way should be well mixed with water, so that it doesn't remain in the tube, where it might get hard and block it. If there is a liquid preparation of a medication, it can easily be administered through the tube. Check with your doctor/pharmacist about alternative preparations of medications.

Placement of a gastrostomy tube generally takes about half an hour. The procedure can be done with heavy sedation and local anesthesia, or under general anesthesia. Most people experience moderate pain for up to several days after tube placement. This pain can generally be relieved well with pain medication. The insertion procedure is generally performed by a gastroenterologist (stomach specialist). Keeping the patient in the hospital after placing the PEG also teaches the patient/caregiver how to use the PEG.

The PEG tube is cosmetically good. Clothing easily conceals it. Medicare and insurance carriers will cover the cost of the surgery, hospital visit, and family training – but generally only covers the cost of liquid nutrition once food is no longer taken by mouth. Check with your Certificate of Coverage or Case Manager.

**Most PALS who have chosen a feeding tube, consider it a good decision!**

The chapter has available to loan a 32 minute video on feeding tube placement and life afterwards. Call the office if you would like to review it.

### **MANAGING SALIVA IN ALS**

The build-up of saliva is a common problem among people with ALS who have tongue and throat muscle weakness and are not able to automatically swallow properly. This build-up of saliva can cause choking and disrupt sleep. Some people have a build up of drooling, also called sialorrhea. Very sweet and sour foods will produce excessive saliva. Relief may come from home remedies, over-the-counter products, prescription drugs, injections, and in some cases, radiation therapy.

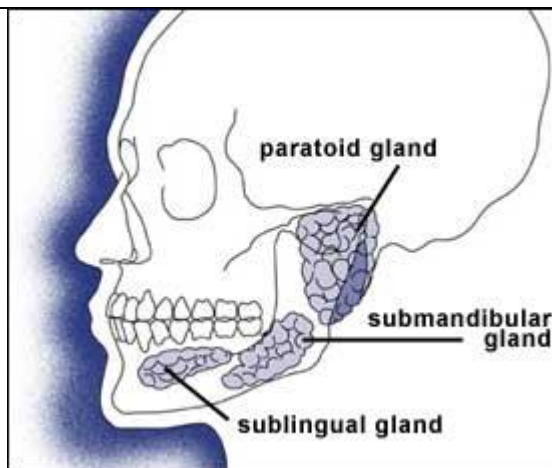
The first step in treating sialorrhea is typically to prescribe medications to reduce the production of saliva. Remember all medications have side effects.

- Antidepressants, such as; Amitriptyline (Elavil), Imipramine (Tofranil) and Clomipramine (Anafranil) have the side effect of dryness of the mouth.
- Scopolamine patch (brand name Scopoderm), which is usually used for motion sickness. The patch is applied to the skin.
- Atropine Sulfate (brand name Sal-Tropine), Clonidine (brand name Catapres) and Propantheline (brand name Pro-Banthine).
- Glycopyrrolate (brand name Robinul).

If medications are not effective, the next step is the injection of botulinum toxin (brand name Botox or Myobloc).

- It works in the same way as the medications, by blocking acetylcholine release from nerve endings, but only at or near where it's injected. Typically the botulinum toxin is injected into each parotid gland. The parotids are the major glands for the thin, watery part of

the saliva. The problem with botulinum toxin is that if it's injected at the wrong site or if it travels, it can block muscles in the area and increase dysphagia (difficulty swallowing) and dysarthria (difficulty speaking). It takes a week to 10 days for the maximum effect to be seen. In about two weeks, you know whether or not it has worked. If it hasn't, after about three weeks, a little more of the botulinum toxin can be injected on both sides. Depending on the response, the botulinum toxin can be injected a second or third time. Typically saliva production is knocked down by about 50 or 60 percent in about half the patients. With a second or third injection, it's usually effective in about 75 percent of patients.



Three major, paired glands - the parotids, submandibulars and the sublinguals - and numerous minor glands throughout the mouth (not shown) normally produce saliva.

Injecting botulinum toxin (brand name Botox or Myobloc) into the parotid glands is one of a number of strategies used for reducing saliva and controlling drooling in ALS.

In a few patients, radiation of the salivary glands has been used as a last option.

- The idea is to damage the gland and to induce scarring which takes several weeks or months. The idea is not to knock down saliva totally but to decrease it to relieve drooling. The problem with radiation is that it's irreversible.

Some ALS patients may experience problems with the thick mucous or phlegm that accumulates in the back of the throat. This is often exacerbated during or after meals or liquid intake. Coughing it up can be a tiring and arduous process, and take its toll on the patient and the caregiver. It also has a tendency to block airways or make food stick in the throat, so coughing it up is essential. Below are treatment options and prevention measures.

Adequate fluid intake is the first essential step in prevention.

- Sips of cold carbonated drinks, hot tea with lemon, lime juice, apple, pineapple or papaya juice, all available at most supermarkets, may help. (Swab the mouth if the PALS is unable to swallow).
- Meat tenderizer (made from Papaya Root) mixed with a little water can also be used to coat the tongue or can be placed under the tongue.
- 12-hour allergy medications such as Zyrtec, Claritin, Robitussin or Allegra significantly reduce the amount of mucus produced.
- Avoid Dairy Products that contain the protein casein that binds with the saliva and forms thick ropy mucous which is hard to break up and/or swallow.

For PALS who face unresolved problems with phlegm, there are medical devices dedicated to assisting with removal of mucous. Discuss with your physician a suction pump and a cough assist machine. Both of these machines are listed as durable medical equipment and are covered by Medicare – remember to check your certificate of coverage for policy specifics. It's best to order and start using a

suction machine (and Cough Assist) early on, while the ALS patient still has the ability to cough.

### **Suction Machine -- [www.respertise.com](http://www.respertise.com)**

A portable suction pump is a lightweight, compact, aspirator designed for upper respiratory oral pharyngeal and tracheal suction. The portable machines generally have a battery backup, as well as a removable AC cord and many come with a carrying case and a DC converter, which allows you to plug it into a cigarette lighter. It also allows you the luxury of bringing it with you when going out. It is helpful to suction the roof of the mouth, back of the throat (watch out for the uvula, you don't want to suck that up) to reduce the stream of mucous. Most Insurances/Medicare cover a training session by an RN who has respiratory care experience to educate and demonstrate safe and effective airway secretion clearance along with this device.

If you have feelings of phlegm stuck in the throat, ask your practitioners about a French suction catheter (#14) – this is a long and narrow soft and flexible plastic tube. It is designed to reach secretions/phlegm effectively down in the throat. A Yankauer suction tube-often referred, as Tonsil Tip is good for mouth area secretion removal only.

### **Cough Assist Machine - [www.coughassist.com](http://www.coughassist.com)**

The Cough Assist machine (also know as a Coffalator or In-Exsufflator) may be effective for patients with an insufficient ability to cough. It safely and effectively clears retained broncho-pulmonary secretions, reducing the risk of respiratory complications. It gradually applies a positive pressure to the airway, and then rapidly shifts to negative pressure. The rapid shift in pressure produces a high expiratory flow from the lungs, simulating a cough. It is easy to use and can be used with a facemask, mouthpiece or an adapter to patients endotracheal or tracheostomy tube. However, PALS who do use the device will usually also

need a suction pump with appropriate suction catheters to clear phlegm and secretions from the throat and mouth as well. Medicare typically covers this device.

### **Loss of Voice**

Some PALS lose voice volume and others lose voice quality. Assistive Technology devices and Augmentative/Alternative Communication devices can help to make it easier for your thoughts, ideas, wants, and needs to be known. By taking advantage of these devices and techniques, you will not have to rely on another person to speak for you.

Augmentative/Alternative Communication" (AAC) refers to any mode of communication other than speech. There are high end devices that will speak for you, interface with your home computer allowing access for emailing/surfing the web/texting, and also interface directly with your telephone. There are also many forms of low end techniques that include: sign language, symbol or picture boards, and easy write boards.

The way a person accesses a device is an important part of communication too. For example, one individual may use fingers for typing, with another moving the eyes to point at letters, thereby spelling words. An access method for an individual will often change as the disease progressively leads to weakening of different areas of the body. This means that a patient who at one point uses hands for communication may lose this ability and eventually have to use another access method. It is important for each individual to know that communication does not have to stop simply because hands become weak and typing is no longer possible.

There are many myths about communication devices. There is no perfect device and no matter how sophisticated the device, it is still more time-consuming than speaking. Having a communication device does not mean you must stop speaking. In fact, you should begin thinking about communication devices well before you lose the ability to speak, because getting the initial assessment, securing the appropriate device, and learning how to use it may take several months. Continue speaking but utilize AAC equipment when faced with an unfamiliar situation such as, a restaurant setting or a large family gathering. When your caregiver and family members cannot understand your speech, a device must be utilized to convey wants, thoughts, and needs.

Your ALSA case manager and the network of ALS experienced evaluators can work directly with you and your family so that communication is manageable throughout your journey of ALS. Section 4 includes a listing of these evaluators throughout Eastern Missouri as well as Central and Southern Illinois.

### **Assistance for Breathing Difficulties**

Pulmonary health is an important issue. It is important for ALS patients and caregivers to learn about mechanical ventilation – both invasive and noninvasive choices - by getting good information and talking to peers. Then when necessary, a person can make an informed decision.

The primary functions of the respiratory system are to bring oxygen into the lungs, transfer the oxygen to the blood, to expel the waste product called carbon dioxide, and to help regulate acid-base balance. Because the lungs have no muscles of their own, the work of breathing is done primarily by the diaphragm and, to a lesser extent, by the intercostals muscles (between the ribs).

Sleep aggravates weak breathing muscles and developing respiratory failure. Thus more breathing problems occur at night. During REM sleep (deep “rapid eye movement” sleep), accessory breathing muscles and other voluntary muscles become so relaxed they are sometimes referred to as paralyzed. That is why breathing problems are first noticed during REM sleep because at that time breathing becomes completely dependent upon the patient's diaphragm, which may be weak due to ALS. This is usually tested at a sleep lab in an overnight sleep apnea study. If daytime breathing tests are within normal range, a simple over-night oximetry study can be done at home (this continuously records pulse and oxygen saturation).

**If your answer is “Yes” to any of these questions, then it is probably time to discuss with your doctor options for breathing support. Notify your physician if you experience any of these.**

- Are you experiencing increased fatigue?
- Do you frequently awaken with a headache?
- Are you having problems sleeping?
- Do you need to use additional pillows when sleeping?
- Do you sleep better in a recliner or chair than in your bed?
- Do you have a poor cough or difficulty clearing secretions?

It is critical to have an effective cough. The strength of a person's cough can be assessed with a Peak Flow Meter. The individual takes a deep breath and then coughs forcefully into the Peak Flow Meter. A “peak cough flow” reading of less than 180 liters/min. indicates a weak cough, according to John R. Bach, MD. A cough can be improved by:

- Use huff coughing (repeated gentler huffs to bring up secretions)



- Manual cough assist methods (having someone push on your upper abdomen as you cough)
- Devices to improve cough and clearance of secretions, [www.coughassist.com](http://www.coughassist.com)

The ability of the lungs to expand should be maintained.

- Use a chest inflation machine
- Use glossopharyngeal breathing (GPB) – sometimes called “frog breathing” (Deep breaths)

Get Adequate Nutrition.

- Unless your muscles get the required amount of nutrition as well as oxygen, they will function less effectively and any weakness will become increasingly noticeable.

Maintain a calm relaxed state

- Panic attacks are common when a person is having difficulty breathing.

If a person needs to use more pillows to elevate his head in bed, or prefers to sleep in a chair or a recliner, or his legs begin to swell, these symptoms may be enough motivation to begin using ventilatory equipment. Physiologic tests should confirm that chronic respiratory failure is developing.

**An ALS patient who is having breathing problems has three choices:**

1. Decline all assisted ventilation and use only palliative care.
  - Discuss Hospice with your physician when vital capacity is 50%
  - Hospice enrollment is typically assigned when vital capacity is at 30%.  
However, other factors play an important role in this decision

## 2. Use only non-invasive Nasal Positive Pressure Ventilation

Noninvasive ventilation provides ventilatory support to a patient through the upper airways. It enhances the breathing process by giving the patient a mixture of air and oxygen from a flow generator through a tightly fitted facial or nasal mask. Also known as Noninvasive Positive Pressure Ventilation (NIPPV), noninvasive ventilation assists the patient in taking a full breath and helps to maintain an adequate oxygen supply to the body.

When these ventilation devices are working properly the carbon dioxide (CO<sub>2</sub>) is eliminated and the patient's oxygen level is high; with improper ventilation the patient's oxygen level drops. If a person is not getting enough breathing support the CO<sub>2</sub> level will climb and leave less room in the lungs for oxygen, so the O<sub>2</sub> will decline.

There are many benefits from Nasal Positive Pressure Ventilation:

- It provides muscle rest and recovery at night.
- It helps reset the CO<sub>2</sub> sensitivity in the respiratory control system.
- It probably improves respiratory mechanics.
- It helps with periods of low oxygen and helps prevent acidosis.
- It improves the quality of sleep.
- It can improve cough and swallowing.
- It improves long-term survival.

Traditionally, NIPPV devices have been for short term use only and a stepping stone to Invasive Home Ventilation. In 2009, technology supported the invention of a new device that is versatile, lightweight, and streamlines the ventilation process with interchangeable active and passive exhalation ports and the flexibility to choose the best available circuit and patient interface. It can accommodate a mask, mouthpiece or tracheostomy. Patients can remain active as possible while

using this type of ventilation to support their breathing. The FDA has approved several types of portable home NIPPV ventilators (e.g. Trilogy™, Newport®, Vela®, IVent, Puritan™, and LTV®) to provide continuous or intermittent positive pressure ventilation through invasive or non-invasive interface.

3. Use whatever ventilation support is needed for long-term survival, including trach positive pressure ventilation. A tracheostomy should be considered if NPPV fails, if the person has a problem with secretions and/or prefers a trach, or when long-term survival is important. Safety may be better with a tracheotomy when daily 18 to 24-hour ventilator life support is needed. If you have a trach, you should also have a good suction machine. More suctioning may be required during the first year while the trach heals and matures. Ventilators are made in portable unit and many people can maintain an active life for many years.

Choosing Home Mechanical Ventilation (HMV) is conditional upon:

- Quality of life is worthwhile.
- Physician presents the options.
- Resources are available (particularly 24 hour caregiving assistance).
- Social support is available for HMV (hand in hand with medical support).
- Insurance coverage for procedure and supplies
- Benefits outweigh the burdens.

In the ALS Care Database Research Study, 66% of participating patients had emergency intubation and 50% of those 66% wished they hadn't made the decision to be vented. It is a difficult process once you are on a ventilator and

wish to wean off of it. You should consult an attorney, hospice organizations, and your physician to discuss the healthcare legalities of this process. Discuss all options with your physician and caregiver to make an informed decision. Make sure your wishes are outlined in an Advance Directives Document (available through the chapter) and your caregivers and family know of this decision. This will allow you to control your own healthcare. It will also put into place a plan of action for your family to follow which will reduce fear for your caregiver as you near those decisions in your journey with ALS.

In 2011, the U. S. Food and Drug Administration approved NeuRx Diaphragm Pacing System for treating amyotrophic lateral sclerosis (ALS) patients who have stimlatable diaphragms and are experiencing chronic hypoventilation.

The FDA Humanitarian Device Exemption (HDE) marketing approval is based on demonstration that NeuRx DPS® could help ALS patients live longer and sleep better than the current standard of care, alone. These findings are the result of a multi-center clinical trial that enrolled 106 patients and treated 86 for chronic hypoventilation.

As the phrenic nerve to the diaphragm muscles fails, patients lose the ability to breathe without ventilator support. In ALS, NeuRx DPS® is implanted through minimally invasive laparoscopic surgery and provides electrical stimulation to the diaphragm muscles. Repeated use of NeuRx DPS® conditions the diaphragm muscles, delaying respiratory failure and the need for tracheostomy and mechanical ventilation.

NeuRx DPS® is a four-channel, battery-powered external pulse generator (EPG) with electrodes that are implanted through minimally invasive laparoscopic surgery. The device provides electrical stimulation to the muscle and nerves of the diaphragm.

During the procedure, a surgeon creates four dime-sized holes in the abdomen and inserts a laparoscope so the diaphragm muscle can be seen. The surgeon then places small electrodes in the diaphragm. The electrodes are attached to the EPG, which stimulates the diaphragm, causing a contraction of muscle. The ALS surgery can be done on an outpatient basis. Post-operatively, the EPG is programmed, and the patients and caregivers are trained on the use of NeuRx DPS®. The stimulation is then used to condition the diaphragm, enabling the patient to breathe longer without the need for tracheostomy ventilation.

## **Multi-Disciplinary ALS Clinics in Eastern Missouri, Central and Southern Illinois and Boarding States**

### **Kansas**

#### **University of Kansas Medical Center**

An ALS Association Certified Center of Excellence  
Department of Neurology - 1008 Wescoe  
3599 Rainbow Blvd./MS 2012  
Kansas City, KS 66160  
Medical Director - Richard J. Barohn, M.D. / April McVey, MD  
Phone: 913-588-0656

### **Indiana**

#### **Indiana University ALS Center at IU Health**

An ALS Association Certified Center of Excellence  
355 West 16th Street, Ste 4700  
Indianapolis, IN 46202  
Medical Director - Robert Pascuzzi, MD  
Phone: 317-963-7385

### **Missouri**

#### **St. Louis University Health Sciences Center**

An ALSA Certified Center<sup>SM</sup>  
Department of Neurology; Neuromuscular Clinic  
3635 Vista Avenue at Grand Blvd.  
St. Louis, MO 63110  
Medical Director: Ghazala Hayat, M.D.  
314-577-8026

#### **St. Francis Medical Center**

Multidisciplinary Clinic  
211 St. Francis Dr,  
Cape Girardeau MO 63701  
Medical Director, Robert Gardner, Jr, M.D.  
573-331-5520

#### **Ozark Medical Center**

1100 Kentucky Avenue  
West Plains, MO 65775

Medical Director - Dr. Vikas Kumar  
Phone: 417-257-6777

### **Kentucky**

#### **University of Kentucky**

An ALS Association Certified Center of Excellence  
KY Neurosciences Institute  
740 South Limestone St.  
Lexington, KY 40536-0284  
Medical Director - Edward J. Kasarskis, MD, PhD  
Phone: 859-218-5061

### **Arkansas**

#### **UAMS ALS/MDA Clinic**

4301 West Markham Street, Suite 500  
Little Rock, AR 72205  
Medical Director - Dr. Stacy Rudnicki  
Phone: 501-686-5135

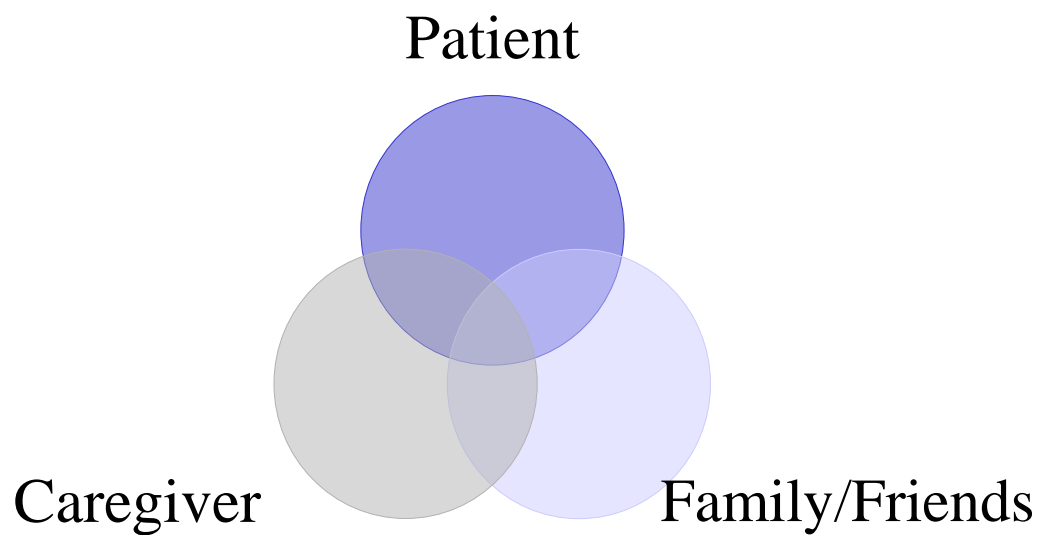
### **ALSA Network of Chapters**

Geography plays an important part in the service delivery. Nationwide ALSA 41 chartered chapters, 1 national chapter, 1 chapter in organization and 1 partner affiliate that work together to help patients across state lines. Each chapter provides similar resources and can assist with vacation travel or clinic visits.

Because of geography, a patient may live in one chapter's service area and attend a clinic in another chapter's service area. We work closely with all ALSA chapters to form a team nationwide.

A detailed listing of ALSA chapters can be found at [www.alsa.org](http://www.alsa.org).

# FAMILY ISSUES





Neuromuscular disease by its very nature means loss. Whether you are the patient, the caregiver, or the friend/family member you will experience grief as functional loss happens. As muscle tissue degenerates, whether quickly or over decades, people undergo a series of losses, not only physical with the loss of strength or the ability to walk or speak, but lifestyle losses as well with changes in work habits, the inability to live alone, or help with daily care. The changes that ALS will bring about are different in everyone's life; however, what can be generalized about ALS is that we grieve after each loss, no matter how large or small. You may not identify your feelings about the losses associated with progressive disease as grief. You may have anticipated changes ahead of time and adjusted your life accordingly, with a minimum of obvious emotions. But even when changes seem to go smoothly, every major loss has an impact. A loss by neuromuscular disease can be like losing a part of yourself or your identity. You may feel you no longer know who you are, but to survive, you'll need to redefine yourself. That redefinition is accomplished by grieving – acknowledging and experiencing whatever emotions arise. Completing the grief process allows you to shift priorities and focus on different aspects of yourself. And with that shift, loss can bring gain – a new way to see yourself and your place in the world. Morrie Schwartz, (ALS patient, Tuesdays with Morrie) stated in his book, Letting Go “ALS is only a part of me, not all of me”. Grieving is as individualized as your course of ALS. There is no formula that works for everyone. Search until you find the ways to pay attention and release the grief that you are experiencing. If you feel stuck or burdened with an unproductive degree of unacknowledged grief or emotion that won't let you go, get connected with a support group or with an individual counselor that knows about neuromuscular illness or specializes in grief. Unresolved grief can lead to anger, resentment, and depression.

Because depression can slow body movement, producing fatigue (similar to symptoms of ALS), signs of depression may be overlooked. If undetected, depression will remain unmanaged, compounding life's misery. Furthermore,

depression is like a contagious disease, infecting an entire household. By recognizing signs of depression, strategies can be implemented for alleviating unbearable grief, enhancing the ability to adapt to losses.

Common signs of depression include: loss of energy, disinterest in activities, lack of desire to interact with family and friends, wishing not to be bothered, crying spells, slowed thinking, forgetfulness, difficulty concentrating and making decisions, sleep disturbances, unkempt appearance and loss of appetite.

Signs of severe depression include: staring into space for long periods, no desire to do anything, communicating only to answer a question, poor concentration or memory, confusion, refusal to eat, weight loss, gastric disturbances, difficulty sleeping or excessive sleep, feeling that there is no reason to go on living and thoughts of suicide. Strategies for maintaining depression include:

1. Express your feelings. Talking about losses is very important in working through grief precipitated by loss. If speech is impaired, find an effective means of communication.
2. Focus on your abilities rather than your disabilities. Use assistive devices to enhance mobility. Do not resist a wheelchair if needed. Modify your home environment for easier accessibility.
3. Set new goals that are achievable. Maintain a purpose in living, despite physical limitations.
4. Have something to do and to look forward to everyday. Keep your mind busy. Plan for pleasurable experiences and achievements.
5. Maintain social interaction with others. Avoid loneliness. Plan enjoyable activities with family or friends. Find accessible methods for leaving your home, going in and out of your vehicle, and visiting others. If traveling is difficult, encourage family and friends to visit with you at home.
6. Maintain control of your life and decision-making. Being in control reduces feelings of powerlessness and helps to prevent giving up.

7. Strive to maintain a positive self-concept. Getting dressed daily and looking one's best can help people to overcome a negative self-image.
8. Seek spiritual care. Faith in God, scriptural meditation and prayer provide peace of mind, joy and hope for tomorrow, despite trials and tribulations.
9. Signs of unresolved depression may warrant treatment or professional counseling. Seek help if needed. By understanding strategies that may assist in adjusting to loss, people with ALS may help overcome agonizing grief and depression. The goal is to change from focusing on what was lost in the past to focusing on what is here to enjoy now and the days ahead.

Providing care for an ALS patient is a major time commitment and an extremely important factor in helping the patient cope with the disease. It is also one of the most heroic acts of kindness and love, one can give to another. The well-being of the spouse or caregiver is also a concern. Because of the progressive nature of the ALS, families continually must find effective ways to cope with the symptoms of the disease. The lack of a set pattern and lack of specific manner of progression make for frustrating and difficult times. Caregivers most often provide care day and night, over weekends and on demand – providing personal care, assistance with mobility, transportation, housework and grocery shopping alone with looking after other family member's needs as well. Caregivers are often employed outside the home and may be the primary source of household income which adds even more demands, responsibilities and stress. "Caregiver burnout" is a problem often faced by ALS families. Feelings of tiredness and frustration are normal to experience accompanied by guilt. Caregivers must learn the importance of having time (respite care) to fulfill their own needs. It is very common to fall into the superhuman mode and try to do everything by oneself. Caregivers must accept help and divide tasks that can be delegated to other family members and/or friends that ask.

No two people with ALS are living in the same situation and no two ALS illness courses will exact the same toll. But all couples, where one partner is

placed in the dual roles of loving partner and personal care provider will experience the greatest test on a relationship and that of their commitment to it. It is essential that couples continue to function as partners, and not become only patient and caregiver. The relationship will be changed forever but it is important to accentuate the positives of this journey and take advantage of the tools that can help preserve this relationship.

With ALS physical changes, comes a shift in the balance of power that is naturally created in all relationships. As one partner becomes dependent, the other must “pick up the slack” to make sure that the household functions, the kids are raised and/or cared for, bills get paid, and provide physical care and emotional support for their loved one. The well partner often needs some extra emotional support to accomplish all of this, but some caregivers are unable to turn to this person to meet this need.

Each partner is also grieving, not only for the life of the person diagnosed with ALS, but with each lost physical task also points to lost dreams and loss in identities. It also brings stress of the changing relationship and added tasks for caregiving duties to the forefront again.

A loss of privacy is also present to both the ill partner and the well partner. A loss in intimacy can develop if both partners do not do everything they can to honor the choice they have made to travel this journey together as a team. The biggest mistake that a person with ALS can do to destroy this relationship is to live with the thought my partner can do it alone or my partner is the only one I will let help me. Other misconceptions include: not paying attention to your partner and asking them when they need help - not when you think they need help or making the decision for them. Although this decision protects the sick spouse, it can and in many instances, directly harm the well spouse both physically and emotionally as well as destroy the bonds of marriage.

Many past caregivers report feelings of anger, guilt, fear, isolation, grief, and financial threat whether they are full-time caregivers or whether their partners have only moderately disabling illnesses. Accompanying those feelings are signs of depression, loss of appetite, sleeplessness, loss of sexual drive, and crying. The act of caregiving itself seemed to bring stress, nervousness, and depression with it. Past caregivers also reported that maintaining the intimacy levels and fulfilling the sexual attention was described as work, rather than loving acts between two people.

Each caregiver will react differently to the added responsibilities, however, at some time during the course of ALS, whether short and rapid progression or a long slow disease course, the changes and grieving can force a shift in the relationship. Both the sick spouse and the well spouse must be cared for! Below are some tips for preserving the relationship: (These tips work well for any relationship, not only for spouses, but for parent/child, sibling/sibling, friend/friend as well).

1. **Maintain equality** within the relationship and find ways for both partners to make significant and meaningful contributions. Take advantage of the resources available to you. The power of technology can allow a person with very little controlled muscle movement to become an active member of the family.
2. Keep **communication** open through family meetings and make decisions as a team. Remember, to try and end the ALS journey as you started it, a team fighting together.
3. **Focus on your relationship:** Research indicates that the better you feel about your relationship with the person receiving care, the less stress you will have. Talk with him or her. Get counseling. If there is serious conflict, invite a third person—one you both know and trust—to help mediate. The results can be

gratifying: spouses with the highest morale generally attribute it to the continuing companionship and good relationship they have with their partners, which can also help sex lives to stay healthy.

4. **Get help!** Take advantage of resources available – especially **respite care**. This allows caregivers to get away from relentless and potentially overwhelming responsibilities for an hour, a day or several weeks by having skilled care personnel stay in the home, or by having their partner stay in a facility which provides an appropriate level of care. Most caregivers report that providing personal care for their spouse takes away from the sexual intimacy needed to maintain strong bonds of marriage.

5. **Stay Independent and Avoid Isolation:** Keep in touch with friends, have people over/visit them. Going out independently will help decrease isolation and foster independence. Not many married couples, independent of ALS, spend 24 hours together and do all activities together

6. **Pay attention** to each other's health, both physical and emotional. Honor each other's need to manage stress, relieve frustration and grieve by taking enough time for themselves.

7. **Manage Stress** – Stress can cause physical and emotional exhaustion. Its symptoms include diminishing self-esteem, a negative attitude, a loss of concern for others, and a loss of focus on your own life. If let unattended for caregivers, it can lead to burnout - It's real and it happens!

8. **Get Your Finances in Order:** Regardless of how little or how much you have, get some help sorting through insurance policies, retirement programs, social security and other government entitlements to find out what there really is to draw on. Keep in mind that specific benefits and programs change from year to year, so re-check periodically. This will help to minimize endless worry about financial strain.

**9. Accept Help:** Don't try to manage ALS all alone. Taking advantage of the offers of help that come from family and friends, can make time to be set aside as couple time. It is important to continue dating activities, or those activities celebrating your love for each other that were in place prior to ALS.

You may not be able to do anything about ALS being in your life, but you control how it impacts your time, energy, attitude and quality of life. Finding ways to acknowledge and nurture your personal life will bring new energy and enthusiasm into your life as a partner, patient, friend or caregiver. Even the strongest individuals, and marriages may need extra concern and help along the path of ALS.

Crisis, whether it be ALS or caring for someone with ALS, tends to heighten our awareness of the spirituality forming who we are. By providing a safe space for spirituality to surface and create sufficient time and a listening presence, an adjustment can be made within ourselves to understand life and death and the choices that lie in front of us better. Spirituality often sounds like it's far away or hard to understand or surely about somebody else.

Spirituality can be confused with religion. Religion practice is a central piece of many people's spirituality but not everyone's. Whoever we are, by virtue of the fact that we are living, breathing people here, we each have our own spirituality. Spirituality's threads include the sounding of our voice/our story, the web of our relationships (self, others, a sense of the divine), and the meaning we make of life as we live it. When we're busy, healthy, or focused on success and goals, spirituality may run quietly, even unnoticed, underneath the surface of our lives. Again, it is important to address these issues to live longer and stronger with ALS in your life. ALS can affect both the patient and the caregiver spirituality.

As frightening and challenging as the diagnosis of ALS is, it is recommended that you share your diagnosis with your family and friends as they will be your support system throughout the course of your illness. Even though, you may not know what words to use or how in-depth your explanation should be. Telling other adult family members and friends may be difficult enough, but finding the words to tell your children is often even harder.

As parents and grandparents, a natural tendency is to protect or shield children from the worries and fears of a serious illness in the family. However, do not be fooled into thinking that your children do not know something is wrong, even if you have not told them. Even the youngest child has the uncanny knack of sensing or figuring out that something is seriously wrong. If you have not opened the dialogue about ALS with your child, he/she may not feel comfortable asking or talking with you about what is going on. Without this conversation, your child can live with fears and worries of the unknown. A child's creative imagination can often conger up scenarios that are grossly inaccurate or terribly frightening. Children can even feel they are somehow to blame for what is wrong and live with unfounded guilt. Therefore, **honesty** is definitely the best policy when sharing your medical situation with children.

In addition to alleviating children's fears, honesty about ALS also promotes the notion that ALS is not something about which the child should be embarrassed or ashamed. As new changes develop in your health and abilities, allowing the child to understand and discuss these changes gives him/her a greater sense of trust in you. The only thing scarier for a child than having a parent who is seriously ill is not having a **sense of trust and comfort** with you, their primary source of security.



Speaking to your child in an **age appropriate** manner is equally as important as being honest about your disease. Not only do children learn and absorb information differently than adults, but they process information differently based upon several developmental stages in their childhood. Intellectually and emotionally, a preschool age child cannot comprehend what an elementary school child can. The same is true for older age groups; their ability to understand and process information and concepts hinges on their developmental maturity. Therefore, the language you use to explain your disease and the amount of information you provide should be determined by the age of your child. If you have children of varying developmental ages, you should be discussing your diagnosis differently with each child. The key is to provide each of them with information that is age appropriate, while introducing a dialogue for future discussion and building an atmosphere of trust and openness.

In talking with a child, use words that a child can comprehend. Clarify your child's understanding as you discuss ALS and its impact on you and your family. It is not uncommon for children to misinterpret what you are saying, simply because they did not understand an explanation or comment you have made. Children may also feel guilty, thinking that in some way they have caused your disease. It is important to reassure them that while doctors do not know exactly what causes ALS, we do know it was not caused by anything you or your child has done or said.

Encourage your children to ask questions and respond as honestly as you can. However, consider the meaning of their inquiries when responding to them. For example, if you should fall and your young child asks, "Are you okay?," s/he is probably wanting to know if you are hurt by your mishap. An older child, however, is more likely to be associating your fall with your diagnosis and may be asking if ALS will continue to affect you in this way. Gauge the complexity of your answer to the age of your child and the context of the question. Remember

that children often learn in smaller “bites” of information, so tailor your responses accordingly. For example, when asked by your child if you will “get better,” you may need to differentiate between a reply about symptom management versus a disease cure. For a younger child, you may want to explain that through managing and accommodating your symptoms, you can move, breathe, or speak “better.” However, for an older child or a child who has more knowledge about your illness, you may need to directly address the fact that there is no cure to stop the disease or reverse the effects it has had on you.

Besides establishing an environment of security and trust, sharing information about your ALS diagnosis should also convey several other messages to your children. Through your honesty and openness, your children/grandchildren will have the basic tools for helping live well together as a family touched by ALS.

- Be willing to admit that ALS is a **confusing and upsetting disease**. Explain that it is hard to understand why ALS happens and what effects it will have on you and your family. Acknowledge your frustration as well as theirs.
- Emphasize that **no one is at fault** for your getting ALS. Explain that doctors and scientists do not know exactly what causes ALS. Especially point out that nothing you and anyone else did caused your ALS. ALS is not a punishment.
- Stress **acceptance and respect** for yourself (or the family member with ALS). Encourage your children to ask questions about this disease and its impact on your life. Also, support their efforts to offer help at what ever level they are able.
- Offer **optimism and hope**. Researchers are working every day to find a treatment and/or a cure. They may find positive results at any time.
  - Also, be willing to “live in the moment.” Allow yourself and your family to enjoy the joys of each day as they occur. This perspective

offers continual hope for “good” days together as you live with ALS.

You should also be aware that your children’s reaction may be different than you expect. For instance, your child may seem disinterested, or ask questions that seem irrelevant. He/she may abruptly begin playing with toys during a conversation or act out in his/her play. Children may demonstrate a range of emotions, initially being angry or sad, but moving quickly into silliness or happiness. All of these reactions are normal for children. It is important to understand that they are processing as is appropriate for children their developmental age. Allow your children time to process the information and work through these reactions. Expression of their emotions and feelings is healthy. Your children need to be allowed to accommodate these changes into their lives, developing the coping skills that will help them, while allowing for on-going openness and dialogue about the changes that are occurring.

As your disease progresses, you will want to keep the lines of communication open and help your child understand and prepare for what is happening. Please consider the following issues:

- Explain changes in your health as they occur in age appropriate terms that your child will understand.
- Do not assume that your child will react to each of your medical or physical changes. Reassess what your child is thinking and feeling. S/he may not be affected by changes as you think they might; alternatively, s/he may be greatly affected by something that you consider minor.
- Validate their feelings as normal. Every emotion they have can be “normal.” Help them find positive ways to express these emotions.
- Recognize that, as your medical condition changes, so will your physical appearance. Acknowledge with your child the social reactions you get and

help them to understand that others may react negatively or rudely due to being fearful or uninformed about a disease they do not understand.

- Talk with your child about issues other than ALS. Ask questions about school, friends, hobbies, television shows, or internet games. Remain an active part of their lives.
- Explain to your child that your irritability or personality changes may be due to the progression of your ALS. Watch for signs of anxiety or feelings of discouragement from your child; they may be feeling that they are to blame for your negative moods.

**The following guidelines will help you talk to your child using language and concepts that are understandable to his/her age.**

## **INFANTS**

Infants are able to sense the emotions of their parents and other caregivers. Although not verbal yet, they may still express their discomfort or concern through their actions. Parents should make every effort to maintain a regular routine and provide nurturance and attention to a child in this age group, when dealing with their ALS diagnosis.

## **PRESCHOOLERS**

Use simple words and phrases in speaking honestly to a preschool-age child. Be sure to reassure them and provide continued attention and emotional support as well as a regular routine, when possible. This environment establishes a sense of security for them. Since preschool-age children have not yet developed their vocabulary, they tend to act out their feelings in play. Some children may act out aggressively on a short-term basis and this should be accepted, as long as they are not hurting themselves or others. Some children may seem to grasp the gravity of the situation, while others this age may seem totally unaffected. Allow

preschoolers to experience their varied reactions while providing them with a safe haven to do so. Avoid punishment, but provide support and understanding. Suggestions: Dolls, puppets, simple books, and stories may aid in talking with this age group. Explain in simple terms what some of your limitations may be due to your illness.

## **ELEMENTARY SCHOOL AGE**

This age group has developed more cognitive skills and will have a better understanding of the significance of your illness. However, coping skills are not very well defined yet, so denial is often used by this age group. Although more verbal than preschoolers, grade school children also tend to express their feelings through their actions. Therefore, you should try to help them find constructive ways to express their feelings and vent their frustrations. It is still important to set appropriate boundaries for this age group, but let them know you are aware of the additional stress they are experiencing. Again, avoid punishment, but communicate your support and understanding through your words and actions.

Suggestions: Write down the name of the disease and use age-appropriate web sites for additional exposure and education. Inform your child's teacher, school counselor, principal, and /or clergyman about your diagnosis and what your child knows; use them as team members in supporting your child. Provide an overview of how your doctors are helping you manage your symptoms; this provides a sense of control to this age child. Give the child concrete ways s/he can help you, i.e., by drawing a picture for you, feeding the dog, or getting the paper.

## **MIDDLE SCHOOL**

Adolescents are already in a volatile, emotional developmental stage without adding the stress of a serious illness of a parent or grandparent. This age group is busy trying to establish their own sense of self and pushing their boundaries, while separating from their parents. This situation makes the adolescent even

more vulnerable, as they struggle between child-like emotions and feelings of being grown up. It is common for children this age to withdraw or act out; this behavior should be tolerated with some parameters. First, you should leave the lines of communication open; acknowledge to the child that while you do not approve of the behavior, you want to support his/her feelings or worries. Offer to provide the child with additional options, such as a professional counselor, support group, or another trusted adult, if s/he feels that would be helpful. These actions show the child that you are giving permission for them to work through their emotions and concerns.

Suggestion: Make your child's school team aware of your child's understanding of your illness. Enlist their help in supporting him/her at school as appropriate. Decide how your family will handle telling your child's friends and/or their families. Provide an overview of how your doctors are helping you manage your symptoms.

## **TEENAGERS**

The teenager is able to think abstractly and understands the illness much like an adult. This age group may still have reactions similar to the middle school age child, depending on their maturity and development. Teenagers need to also know the lines of communication remain open and that they have permission to voice feelings and concerns. Anger is a normal reaction for this age group, but this child may react in any number of ways. Be sure this child has someone outside of the household with whom they can talk. While this person might be an adult family friend, a school counselor, or a professional counselor, s/he should be someone who your child trusts and who will keep conversations confidential. This age child may try to take on an adult role, so be careful not to expect him/her to act as chaperone for you. This situation creates too much responsibility and forces the child into dual roles with you, which is too emotionally burdensome and confusing. Encourage this child to help out if they

want to, but do not expect them to take on the primary caregiver's role or other additional duties.

Suggestions: Answer questions as honestly and completely as possible. Encourage this child to do additional research on-line or in the library, being sure to recommend reliable resources.

## **YOUNG ADULTS/COLLEGE AGE**

The young adult child faces many of the same issues as the older teenager. While this child will comprehend, react, and be able to assist in much the same ways as an adult, they still must process their worries and emotions of having a parent or grandparent with a life shortening illness. Additionally, this child may be living outside of the home, perhaps away at college or already supporting themselves in their own living accommodations. If this is the case, they may even be living in a different city. These parameters create further emotional burden, as this young adult realizes the implications of the illness and feels the responsibility of helping in some way. It is important to encourage this adult child to seek outside counseling for emotional support.

Suggestions: Most universities offer counseling services through their health services department for free or for a minimal charge. Encourage your college student to utilize this resource. If your young adult child is employed, many health insurance programs also have mental health benefits for counseling. Encourage your young adult to do additional research on ALS for their own education and understanding. Also, several websites focus on the needs of the college-age student or young adults with a seriously ill parent.

The Chapter also has available an ALS Activity Book designed to assist parents and grandparents with the explanation of ALS and the changes that are happening in a child's life. This book is appropriate for ages 5-12.

Some of the most difficult problems that people recently diagnosed with ALS must face can be those associated with work. Depending on the severity of symptoms, deciding when to tell their employers about their situation, estimating how long they will be able to work, evaluating whether they can work part time or from home, wondering how to ask for special workplace modifications, and analyzing the financial impact of forfeiting a full-time or part-time salary all can be serious issues.

There is quite a bit of information and advice available to help and under the Americans with Disability Act, employers must make reasonable accommodations for workers who are faced with a life threatening, chronic, or short term illness. Your ALSA case manager can assist with these accommodations when the time is appropriate.

Gayle Backstrom, who wrote *I'd Rather Be Working: A Step-by-Step Guide to Financial Self-Support for People with Chronic Illness*, suggests that people who are grappling with issues related to their ability to continue to work consider doing a self evaluation that will help them take stock of the demands of the job – physical, mental and emotional – and their own situation physically, financially, emotionally, and mentally. For example, people with ALS should consider:

- What are their physical limitations now?
  - Can they use their fingers and hands or hand to pick up an object, type on a computer keyboard or calculator, operate other equipment?
  - Do they have weaknesses that will limit their ability to get to work and then get to the work or office environment?
  - How much weight can they lift safely?
  - How long can they stand, and how far can they walk?
  - Do they need a cane, walker or wheelchair?



- Do they have trouble maintaining their balance?
  - Can they handle shaving, make-up and getting dressed for work?
- What is their stress level and is it affecting their ability to concentrate and feel motivated?
  - Do they have trouble dealing with the everyday pressures of a work environment?
  - Can they motivate and lead others (if that's a part of their job)?
- What is their emotional perspective?
  - How are they coping with the diagnosis?
  - Have they reached an acceptance of the situation?
  - How is their self confidence?
- What is their situation at work?
  - Does the job create a sense of satisfaction and meaning?
  - Do they have a good relationship with their boss or employer; are they confident that if challenges developed, they could discuss them if needed?
  - Is the work something that could be done at home occasionally, part time or regularly?
- How important is the job to their financial stability?
  - What are the financial pros and cons of staying employed now?
  - Are there milestones which, if reached, would have a significant positive impact on their financial stability (such as a stock option maturing, a bonus or commission being issued, etc)?
  - If their salary were reduced or eliminated, are there other family members who could reasonably return to work to help stabilize the financial situation?

The progressive nature of ALS will require that this self-evaluation occur fairly regularly. Questions related to how and when to disclose the diagnosis of ALS to an employer and what implications that may have on their work situation should be answered by legal counsel or social workers with experience dealing with personnel issues.

Because of the nature of this illness, the diagnosis of ALS means uncertainty. You may feel that there are more unknowns than answers to your numerous questions. As a PALS, you may be concerned about how ALS will affect your family, how you can best prepare them and yourself for the future, or how to get your own affairs in order. As a CALS or other family member, you may want to know how you can support your loved one through their illness as well as meet your own needs for support and guidance in a future without them.

To better assist you with some of these issues and to equip you with resources as you move through uncharted waters, the Chapter offers support with anticipatory grief support and a bereavement program designed to help you and your family make the most of the time you have together. This program encompasses both during and after-care support for you and your loved ones, utilizing numerous resources and tools. The Chapter has professionals available to assist you with specific concerns and needs. Another manual will be presented by your ALSA case manager to introduce these services to your family. Below is an insert from that manual:

As one faces a terminal illness, such as ALS, it can be rewarding and fulfilling to review one's life journey and reminisce about favorite people, experiences, and events. The following activities are intended to provide suggestions for remembering and reflecting on your life, in order to help you celebrate your successes, cherish your loved ones, and honor your journey. These exercises may also allow you to reconcile or accommodate difficult or painful memories or

events, by putting them into the perspective of your history and growth, while providing you with the opportunity to forgive yourself and others as is appropriate.

**Life Review Timeline:** Place the major events of your life on a timeline. You may need several pieces of paper taped together or a large poster board to accommodate the length of your timeline. Measure off periods of time, such as every five or ten years, to give yourself a graph on which to work. Include your successes, special dates, important events, and favorite memories. If you are willing, allow other family members and friends to contribute their memories of you, too. This activity allows you to acknowledge your accomplishments, reflect on troublesome or painful events, and recognize special people or times throughout your life. This timeline, created by you, will also serve as a wonderful keepsake for your family.

**Count Your Blessings:** Make a list of all the things you are thankful for, such as loved ones, friends, possessions, special events, honors, successes, and abilities. Whenever you need a “pick me up,” read through your list and reflect on all the things that you are thankful for. Another option is to keep a “Blessings Journal” by writing five things you are thankful for at the end of each day. Some days your lists may look the same; other days you may find new treasures to be thankful for. By focusing and considering all these positive influences, you are celebrating the life you have created and honoring the blessings that have been given to you.

**Messages to Loved Ones:** This activity can take several forms, all of which provide your loved ones with tangible messages from you for the future.

- **Video taped messages:** Have someone video tape you as you record messages to your loved ones, to be viewed at a later date. These messages could be your thoughts and wishes on special occasions, such as

birthdays, graduations, or weddings, or they could be general communications that you would like your loved ones to be able to replay and share in the future.

- Cassette recorded messages: You can also record a verbal message, without the visuals of the video, for your loved ones under the same guidelines as above.
- Written letters: Write or type letters to loved ones expressing your thoughts and wishes. You can also dictate your letter to a trusted friend or family member, if you are unable to write. These letters can be sealed and dated to be opened at a time of your designation.
- Readings: Have someone video tape you or record your voice on a cassette tape, as you read a child's favorite story, recite a special poem or prayer, or sing a song.

**All About Me:** Answer any or all of the following questions. Consider these queries as starting points for discussing your life, your dreams, your ambitions, and your reflections with your loved ones. Have someone record or write down your responses to create a treasured collection of your responses.

- Who was your best friend in childhood?
  - Where did you live when you first moved out of your parents' house on your own?
  - Of all the cars you have owned, which was your favorite and why?
  - What is your favorite book?
  - What is your favorite song?
  - What is your favorite movie?
  - What is your favorite food?
  - How did you meet your spouse?
- Talk about your dating/courtship.

Who proposed to whom and how?

- What was your first job?

If you could have been in any occupation, what would it have been and why?

When you were a child, what did you want to be when you grew up?

- What quality do you think is most important in a friend?
- What is your favorite attribute or ability in yourself?
- What was the scariest moment in your life?
- What was the funniest thing to ever happen to you?
- What was the saddest moment or event in your life?
- What was the most unusual thing that ever happened to you?
- If you were stranded on a desert island, which three things or people would you want to have with you?
- If you could change one thing that you did in your life, what would it be?
- What are you proudest of?
- What is your favorite hobby and why?
- What was your favorite subject in school and why?
- Name three people that influenced your life the most.
- Which teacher had the biggest impact on you and why?
- If your life story was made into a movie, who would portray you?
- If a genie could grant you three wishes, what would they be?
- What is your lucky number?
- Describe your greatest adventure.
- Talk about a time when you were awestruck.
- What was your favorite thing about your mother?
- What was your favorite thing about your father?
- What was the nicest thing someone ever did for you?
- What makes you laugh?
- Name three people that you admire and why.

**Journaling:** Keep a journal of your thoughts, concerns, wishes, and feelings. You can purchase a blank bound journal from the book store, use a simple spiral notebook, or type your journal on the computer. If you want your journal to be private, explain this to loved ones who might have access to it. Make arrangements with a trusted family member or friend to protect the journal after your battle with ALS has ended. Instruct them to either discard or destroy the journal, or allow them to share your entries with selected loved ones of your choice. Journaling can be a cathartic way of expressing emotions and releasing pent up feelings. It can also serve as a process of self-reflection, soul-searching, and life celebration. Don't over think what you are writing; allow your thoughts to flow onto the paper without editing or critiquing yourself. This process will allow you to address issues that are concerning you and work through difficult or unknown problems. It will also allow you to document what is important and meaningful to you.

**Recommended Reading on Emotional Aspects of ALS: (These books are available to borrow through the chapter)**

- Caregiving: The Spiritual Journey of Love, Loss, and Renewal by Beth Witrogen McLeod.
- Tuesdays with Morrie by Mitch Albom
- Letting Go: Reflections on Living While Dying by Morrie Schwartz
- Chocolate Kisses by Ann Wade
- Learning To Fall: The Blessings of An Imperfect Life by Philip Simmons
- Transcending Loss: Understanding the Lifelong Impact of Grief and How to Make It Meaningful by Ashley Davis Prend
- Share The Care: How to Organize a Group to Care for Someone Who is Seriously Ill by Cappy Capossela and Sheila Warnock
- Rowing Without Oars: A Memoir of Living and Dying by Ulla Carin Lindquist
- Luckiest Man; The Life and Death of Lou Gehrig by Jonathan Eig

## Morris Schwartz shares his experience in coping with ALS

Morris Schwartz, Ph.D., a professor emeritus of sociology at Brandeis University was diagnosed with ALS in the summer of 1994 and passed away in November, 1995. He devoted his last year of life to using his illness to teach others about living and dying. He was an inspiration to friends, students, and colleagues – young and old – on dying with dignity. He wrote 75 aphorisms that he called “Reflections on maintaining one’s composure while living with a fatal illness.” A book of his aphorisms, called *Letting Go*, was published in June 1996 by Walker & Co., NY.

Here is a portion of observations from Dr. Schwartz based on his experience in coping with ALS.

- Be clear about what you need and want.
- Ask for it.
- Persist in trying to get it.
- If you don’t get it, know when to give up trying and accept the loss.
- Now you can experience more freedom to be who you really are and want to be because you now have nothing to lose.
- Learn how to combine detachment with involvement, caring with distance, despair with hope, life with death.
- Let others’ affection, love, concern, interest, admiration, and respect be enough to keep you composed.
- Be patient with and accept your and others’ shortcomings and limitations –again and again and again.
- When you are utterly frustrated and angry, don’t be afraid to express anger or to curse silently, under your breath, or out loud if the situation permits. You don’t have to be nice all the time and get people to like you and want to be with you – just most of the time.
- If you need to rail against your fate, do so while you also try to use your situation to enhance and transcend yourself and achieve a higher level of functioning, whatever that may be.
- If you can’t have large victories or achievements, be grateful and celebrate small ones. Many small ones may add up to a large one.
- Watch for and enhance that which motivates you to be composed and involved.
- Find ways to accept, come to terms with, and reduce your fear of death.
- Keep your heart open for as long as you can, as wide as you can, for others and especially for yourself. Be generous, kind, decent, and welcoming.

- Be kind and loving to yourself. Befriend yourself. Be compassionate to yourself. Be gentle towards yourself. Do not put yourself down or criticize yourself continuously.
- Friends and family may see you as less incapacitated than you are because they want you to be “better.” They have this need because they care about you. Accept this, while trying to convey your reality without imposing it on them.
- Whenever a lessening of a physical power occurs, it will always feel too soon. Expect this reaction, and perhaps by preparing for it mentally, you can soften its impact.
- Resist the temptation to think of yourself as useless. It will only lead to depression. Find your own ways of being and feeling useful.
- At some point be prepared to deal with profound contradictory feeling – for example, wanting to live and wanting to die, loving others, and hating them.
- Watch out for emotional, spiritual, or behavioral regression when you are tired, sleepless, or anxious. Try to find ways to avoid that state or to get yourself out of it.
- Be aware that you are living in the shadow of increasing dysfunction, discomfort, dependency, and death. Come to terms with this shadow in whatever way you can.
- Allow yourself to be admired – to be an inspiration to others for your behavior, attitudes, and spirit.
- Find a time of the day to face fully your dread, horror, anxiety, fear, rage, and anguish in regard to past, current, and future losses and dysfunctions. Weep and grieve deeply at this time. Once you have honored these feelings, be done with them and go back to your involving life. You may find that you are strengthened by your tears.
- Maintain necessary illusions, but not completely unrealistic ones. For example, to hope that my ALS will reach a plateau or move slowly is realistic, but to expect to be cured is not. Be hopeful, but not too foolishly hopeful.

*Strength does not come from physical capability – it comes from an indomitable will.*

*~ Ghandi*



## Caregiver's Bill of Rights:

I have the right:

To take care of myself. This is not an act of selfishness. It will give me the capability of taking better care of my loved one.

To seek help from others even though my loved ones may object. I recognize the limits of my own endurance and strength.

To maintain facets of my own life that do not include the person I care for, just as I would if he or she were healthy. I know that I do everything that I reasonably can for this person, and I have the right to do some things just for myself.

To get angry, be depressed, and express other difficult feelings occasionally.

To reject any attempts by my loved one (either conscious or unconscious) to manipulate me through guilt, and/or depression.

To receive consideration, affection, forgiveness, and acceptance for what I do, from my loved ones, for as long as I offer these qualities in return.

To take pride in what I am accomplishing and to applaud the courage it has sometimes taken to meet the needs of my loved one.

To protect my individuality and my right to make a life for myself that will sustain me in the time when my loved one no longer need my full-time help.

To expect and demand that as new strides are made in finding resources to aid physically and mentally impaired persons in our country, similar strides will be made towards aiding and supporting caregivers.

# Living with ALS

## Chapter & Community Resources



## **BACKGROUND & HISTORY OF THE ALS ASSOCIATION**

The ALS Association, St. Louis Regional Chapter had its beginnings in 1984 when a local businessman, Ronald Flowers, was diagnosed with ALS. Ron's business partner, R. Scott Davis, scheduled a general meeting of interested individuals and, as a result, a support group was formed. The purpose of the group was to unite efforts in providing ALS information, patient and family support, and fund-raising for programs and research.

In August 1987, this group became affiliated with the National ALS Association beginning as the Greater St. Louis Chapter. In 2001, the chapter adopted a new name (St. Louis Regional Chapter) to reflect its expanded service area of Eastern Missouri as well as Central and Southern Illinois.

In 1992, The Association became a member of the Combined Health Appeal of Greater St. Louis and in 1999 became a member agency of the United Way of Greater St. Louis. The Chapter continually accesses support from these agencies in Eastern Missouri as well as Central and Southern Illinois counterparts.

The Chapter has a solid reputation within the National ALSA Organization with many of its programs being used as templates for other chapters. The St. Louis Regional Chapter has been named Chapter of the Year in 1999, 2001, and 2004 with additional consistent recognition awards in patient and family services, public and professional education, advocacy, and fundraising.

Through the years, the goals of the Chapter remain the same:

- ◆ To provide programs, support, and encouragement to ALS patients and their families.
- ◆ To provide community and healthcare education, and to stimulate public awareness about ALS.
- ◆ To raise funds for local patient services and national research.

A yearly review of the strategic plan by the board of directors guides The Association's efforts every day and includes strategies to lead the staff and board in fulfilling the mission of The Association. The mission reads: *Leading the fight to treat and cure ALS through global research and nationwide advocacy while also empowering people with Lou Gehrig's Disease and their families to live fuller lives by providing them with compassionate care and support.* Our Vision is to create a world without ALS.

In 2001, The Association adopted the chapter tag line of *Where Patients Matter Most* and this focus is consistent throughout all the departments' efforts; Patient and Family Services, Education and Public Policy, and Development.

In July of 2005, the chapter was instrumental in assisting St. Louis University Hospital, Department of Neurology in being certified as a Center of Excellence through The National office of ALSA. This Clinic offers a 10 member multi-disciplinary approach for managing ALS symptoms. In November 2010 an ALS clinic was opened at St. Francis Medical Center. This clinic offers a multi-disciplinary approach for managing ALS symptoms.

The chapter is bound to the medical community through its Medical Advisory Committee. This medical advisory Committee exists to offer guidance and literature review of chapter materials and programs. It is comprised of multi-disciplinary team members of all centers and clinics, which the chapter has community collaboration with. This committee does not meet regularly, however, is available on an as needed basis.

The chapter stays true to its grass roots beginning and is volunteer driven. Lou's Crew is a multifaceted volunteer program that allows volunteers to participate in patient services, public policy, and fundraising efforts with The Chapter. If you

would like to volunteer, please contact the office. Many opportunities exist through committee work, special events, patient projects, and advocacy.

## **CARE AND FAMILY SERVICE PROGRAMS**

### **- *Available to Assist You and Your Family***

Through the Care Services & Programs Department, the chapter offers a comprehensive network of in-home services free of charge to patient and family members. Additionally, a strong alliance exists with other not-for-profit organizations that are familiar with ALS. Lastly, a community partnership program with multiple local service providers offers specialized services to patients and families. Through these collective efforts, the chapter has created a solid continuum of care for ALS patients in Eastern Missouri as well as Central and Southern Illinois.

Through case management, patient services coordinators will meet with your family to assist you to navigate through the health systems, increase functional independence, and help you maintain control of your healthcare throughout your journey with ALS. They will also work to identify other community resources, ALS specific and general healthcare, to support your family as well.

## **IN-HOUSE NETWORK OF SERVICES**

- Eligibility of the services are dependent upon the patients choice of Medical Provider, and access to other community resources.
- *Respite for Caregivers (in home medical care)*
- *Nutritional Supplement Support*
- *Supportive Therapies (Physical Therapy, Occupational Therapy and Speech Therapy visits– including orthotics, braces, unlocking of SGDs for computer access and Bluetooth technology for wheelchairs.*
- *Counseling Services for Patients, Caregivers, and Children*

- *Catchers Fund Flex Grants for Home Accessibility Modifications, Transportation, and Medications Assistance*
- *Durable Medical Equipment, Computer Access, Speech Generating Devices, and Assistive Technology Loan Closets*

In addition, information is available on all aspects of ALS in printed, video, and CD-ROM formats available through the chapter's loan library. Quarterly, the chapter hosts patient education meetings focusing on a topic relevant to ALS. For example, an expo on mobility aids, or an expert lecturer on genetics.

In 2007, the chapter updated a previous educational ALS Activity Book, as a tool for parents to teach their children about ALS. Since that time, many psychological professionals and schools throughout the service area use this as a tool for their clients, students, and families.

### **Share our Strengths Support Network Program**

In 2009, the chapter adopted the Extra Hands program from The Jack Orchard ALS Foundation. The chapter felt it was important to continue to meet the needs of patients and expanded the in home volunteer based program to perform non-medical tasks and chores to assist the patient to remain in their homes and provide relief for the primary caregiver of these tasks.

Assistance is available through the chapter if a family has a network of willing friends or if they need assistance identifying a team of helpers. Every patient family is matched.

### **COMMUNITY CONTINUUM OF CARE**

The Chapter and Clinics partner with many organizations and companies through the

Gold Star Recognition Program. The Gold Star name and logo offers instantaneous assurance that the company excels at service delivery, has a true understanding of ALS, and is dedicated to improving the quality of your life.

- Massage Therapy - see brochure at end of section
- Augmentative Communication and Assistive Technology Assessments
- Prism Medical Ltd, Nightingale Home Modification Program

In addition, the chapter partners with the following durable medical equipment (DME) companies that help manage the chapter's loan closets program. These DME providers specialize in the equipment that is needed for an ALS patient. They work directly with Prism for home modifications as well.

#### Illinois

- NuMotion - Carterville

#### Missouri

- Alliance Rehab & Medical - Sikeston & Ozark
- NuMotion - St. Louis & Cape Girardeau

## **Government Resources for ALS patients**

### **Veterans Benefits for ALS**

Thanks to the efforts of The ALS Association, key members of Congress, advocates and the Department of Veterans Affairs, ALS has been listed as a disease entitled to presumptive service connection. This means that if a service member is diagnosed with ALS his or her condition will be presumed to have occurred during or been aggravated by military service and as such be entitled to service connection and full benefits.

The VA has officially published regulations establishing ALS as a service connected disease. As of September 23, 2008, all veterans with ALS automatically will be eligible for benefits, no matter when or where they served and no matter how soon after discharge they were diagnosed. As of January 2011, VA now grants service connection for ALS at the 100% rate. VA can pay at rates higher than 100% when entitlement to Special Monthly Compensation is established. This includes loss of use of hand, foot, combination of hand, foot, loss of speech, PEG tube, ventilator dependent or assistance with activities of daily living.

### **ASSISTANCE FROM THE PARALYZED VETERANS OF AMERICA (PVA)**

The chapter has been advised that Veterans with ALS should contact their PVA rep for assistance in the changes of benefits.

Paralyzed Veterans of America  
400 South 18th Street  
St. Louis, MO 63103  
314-552-9887



To find the nearest VA Medical Center near you: [www.va.gov](http://www.va.gov) or call the chapter office for a listing of VA Hospitals/Clinics within the service area.

### **SOCIAL SECURITY (SSA), DISABILITY (SSDI), AND MEDICARE BENEFITS FOR ALS**

In 2003, the Federal Government through the tireless effort of ALSA and its lobbyists lifted the two year waiting period for ALS patients to receive Medicare benefits and placed an ALS diagnosis on a presumptive eligibility list for Social Security and Disability benefits. The listing of impairments is a section of SSA's regulations that describes medical conditions that are so severe that SSA deems automatically that a person is disabled. This means that ALS patients do not have to justify or prove their disability status - patients are automatically enrolled as long as work guidelines are met that would make them eligible for Social Security Disability.

SSDI, or Social Security Disability Insurance, offers financial assistance to disabled workers. This program is funded by the income taxes that workers all over the country pay into the system. Eligibility for SSDI is based on an applicant's work history. The SSA assigns a certain amount of "work credits" to each quarter that an individual works and pays taxes. To qualify, an applicant must have earned enough work credits throughout his career. Learn more about SSDI and work credits here: <http://www.disability-benefits-help.org/ssdi/qualify-for-ssdi>.

The second program that offers disability benefits is called Supplemental Security Income (SSI). Eligibility for this program is based on an applicant's financial standing rather than their work history. To qualify, applicants must fall under specific financial limitations and meet the SSA's definition of disability. This program is ideal for disabled individuals who may not have the work

credits to qualify for SSDI. Learn more about qualifying for SSI, here: <http://www.disability-benefits-help.org/ssi/qualify-for-ssi>.

It is important to note that some applicants may qualify for both SSI and SSDI. Also remember that these programs do not take the place of health insurance. However, individuals who qualify for SSDI qualify for Medicare after a two year waiting period. Individuals who qualify for SSI automatically qualify for Medicaid. For more information about Medicare and Medicaid, visit this page: <http://www.disability-benefits-help.org/faq/medicare-vs-medicaid>.

On average, it takes about three months to get a decision. However, this is only an average. Experiences can be shorter or longer. SSA has an expedited procedure for processing terminal illness cases to ensure that a favorable decision can be made as quickly as possible. The code word for this type of case is “TERI” case. A person with ALS, particularly if advanced symptoms are present, should advise SSA of this fact at the time of application and ask to have terminal illness case procedures applied.

## **Medicare**

Since 2000, many changes have gone into effect that increases PALS benefits through Medicare. In 2000, the 24-month waiting period for Medicare coverage of people diagnosed with ALS was lifted. In 2001, Speech Generating Devices were included as standard durable medical equipment for ALS patients. 2002 included changes in the homebound status for disabled individuals and 2003 brought the addition of ALS to the presumptive eligibility listing meaning patients do not have to prove how ALS disables them. In 2006, Medicare initiated prescription drug coverage meaning all persons with ALS who have or are eligible for Medicare will have access to coverage for Rilutek and other medications. Medicare and most private insurances will not cover many devices

that will maximize your functional independence and there can be a large financial expense associated with purchasing these items. **Remember to check with the ALSA loan closet before purchasing an item out of pocket. The chapter ideally keeps these particular items in the loan closets. A CD-ROM is included in the back of this section that explains many of the items.**

Medicare generally covers 80% of costs and becomes a person's primary insurance. It can be combined with a private insurance or a Medigap policy to fill the 'gaps' in the original Medicare Plan.

MEDIGAP - If you are in the Original Medicare Plan and have a Medigap policy, then Medicare will pay its share and your Medigap policy will pay its share of your health care costs.

Currently, there are ten standardized Medigap plans called "A" through "J." Medigap policies must follow Federal and State laws. These laws protect you. The front of a Medigap policy must clearly identify it as "Medicare Supplement Insurance." Each plan, A through J, has a different set of benefits. Plan A covers only the basic (core) benefits. These basic benefits are included in all the Medicare plans (A through J). Medigap Plans B through J offer extra benefits. Plan J offers the most benefits.

PART A - covers inpatient care in hospitals, critical access hospitals, and skilled nursing facilities (NOT custodial or long term care). It also helps to cover the cost of hospice and some home health care.

PART B - covers doctors' services, outpatient hospital care, physical and occupational therapy, home health care, and medical equipment. If you do not enroll in Medicare Part B when you first become eligible, you will face penalties if you enroll at a later date. You will pay increased monthly premiums equal to 10% more for each 12 month period that a person was eligible for Part B, but did not enroll. This penalty will apply for as long as someone has Part B.

Part D - In addition to new coverage for prescription drugs, this coverage also provides additional assistance to those with limited financial resources. In fact, nearly eight million people automatically qualify for this assistance, including people with Medicare and Medicaid, Supplemental Security Income (SSI) and Medicare Savings Program coverage.

Coverage and costs vary depending on which plan is selected and is not automatic - **A person must enroll in PART D and choose one of the plans offered through your state of residence.** Most people will pay a monthly premium (about \$37 in 2006) and an annual deductible (up to \$250 in 2006). For drug costs between \$251 and \$2,250, beneficiaries pay 25% of costs, with Medicare covering 75%. Beneficiaries are responsible for paying all drug costs between \$2,251 and \$5,100. Following that, Medicare will pay 95% of drug costs for the remainder of the year.

The Medicare program's website, [www.medicare.gov](http://www.medicare.gov), includes a section on both government and private programs that offer prescription drug discounts, including Medicare-approved drug discount cards. In addition to providing general information on how the discount cards work and how to determine whether you are eligible to use the cards, the site allows you to compare the discounts offered by various drug cards for the drugs you use most frequently.

Additional information and assistance is available on the Medicare website at [www.medicare.gov](http://www.medicare.gov) and the Medicare hotline, 1-800-MEDICARE. Questions can be directed to local State Health Insurance Assistance Programs (SHIP).

## STATE OF ILLINOIS COMMUNITY RESOURCES FOR PEOPLE WITH DISABILITIES

Medicaid - On August 13, 2009, Public Law 96-0351 Amends the Illinois Public Aid Code. This Provides that the Department of Human Services, in consultation with the Department of Healthcare and Family Services, shall develop a demonstration project within the Home Services Program under which a spouse may be reimbursed for providing care to his or her spouse, who is eligible for services through the Home Services Program and who meets the criteria for the demonstration project. Provides that the demonstration project shall operate in selected counties and be limited to serving no more than 100 unduplicated persons in a State Fiscal Year. Sets forth required components of the demonstration project. Requires that development of the demonstration project begin by July 1, 2009. Requires an interim report to the Governor and the General Assembly by March 1, 2010 and a report of findings and recommendations by March 1, 2011. Effective immediately.

Department of Human Services Division of Rehabilitation Services - provides services to individuals with disabilities under age 60. Financial asset limits apply. Services include personal assistants, electronic home response, home modifications and more.

<http://www.dhs.state.il.us/ors/hsp/> or 800 843-6154.

Illinois State Treasurer's Office Ready Access Program - The State Treasurer's Office works with financial institutions to provide low interest loans for assistive technology and accessibility for individuals with disabilities. Total gross income per household shall not exceed \$100,000 and combined net worth of the household shall not exceed \$500,000. Borrowers can contact one of the participating Ready Access lenders or call the Illinois Treasurer's Office at 312-814-1793. <http://www.state.il.us/treas/Programs/rap.htm>

TechConnect Low Interest Loan Program provided by several state agencies. The Assistive Technology Loan Program provides loans to Illinois residents with

disabilities and/or their families on behalf of the individual with a disability to purchase assistive devices, services and home modifications. Income limits apply. A family of two must not exceed an annual income of \$64,150. (217) 522-7985 or <http://techconnect.iltech.org/>

The UCP Ramp Up Foundation provides grants to low income individuals with disabilities living in a single family home in the state of Illinois. Income limits apply. Funds are used to build ramps, lifts, accessible bathrooms and kitchens, door widening, elevators, stair glides, etc. Call Beth at 708.444-8460, ext. 272 for more information.

AMBUCS (American Business Clubs) is a charitable service organization dedicated to creating mobility and independence for individuals with disabilities. Their services include ramp building. Call the National Office (800) 838-1845 to find your local chapter or visit [www.ambucs.com](http://www.ambucs.com).

The Illinois Department on Aging provides a variety of services to individuals over age 60 and some services to individuals with disabilities. Services include case management, caregiver programs, homemaker services, senior companion services and many more. For additional information or to find your local agency on aging, visit <http://www.state.il.us/aging/> or call 800-252-8966.

**Independent Living Centers** - (CILs) are private, nonprofit corporations that provide services to maximize the independence of individuals with disabilities and the accessibility of the communities they live in. They are based by counties in IL.

- provides a durable equipment loan program

- provides **Assistive Technology Services** help consumers obtain, repair and understand how to use assistive devices such as ramps, wheelchairs, TTY's, and computers.

## **Disabled Parking Rules - State of Illinois**

State law allows for Disability License Plates to be issued to a qualifying applicant, their parent, immediate family member residing in the same household or legal guardian. In addition to the one set of Disability Plates, a parking placard also may be issued in the disabled person's name, which may be used in another vehicle. The Office of the Secretary of State is committed to improving the parking program for person's with disabilities by ensuring that those who qualify for the program have the accessibility they deserve. If you have any questions or need more information, please contact

Persons with Disabilities License Plate/Placard Unit,  
Vehicle Services Department,  
501 S. Second St. Rm 541,  
Springfield, IL 62756,  
(217)782-2709, (217)782-2434 or (217)782-3166.

Effective January 1, 2002, to qualify for the Secretary of State's Parking Program for Person's with Disabilities, an applicant must have one of the following five specific medical conditions:

- Can not walk without the assistance of another person, prosthetic device, wheelchair or other assistive device
- Be restricted by lung disease to such a degree that forced respiratory expiratory is limited
- Must use portable oxygen

- Have Class III or Class IV cardiac condition according to standards set by the American Heart Association
- Be severely limited in the ability too walk due to an arthritic, neurological, or orthopedic condition.

A licensed physician must certify on the application that you have one of the five qualifying disabilities.

Forms available at all senior citizens facilities and Secretary of State Offices, and online at [www.cyberdriveillinois.com](http://www.cyberdriveillinois.com)

- Provide a copy of vehicle registration or title
- Vanity or personalized disability license plates are available
- Placards must be hung from rearview mirrors.
- If current plates are valid for more than 90 day, application for replacement plates fee of \$14.00 is assessed. If current plates expire within 90 days, the annual \$78.00 fee is expected with completed application.

### **STATE OF MO Resources for People with Disabilities**

- Department of Health and Senior Services - Provides home services (homemaker, personal care, respite) for people over age 60 or disabled. Must meet eligibility requirements (financial need). [www.dhss.mo.gov](http://www.dhss.mo.gov) or 800-735-2466
- Community Leaders Assisting the Insured of Missouri: Counseling to seniors and disabled for Medicare insurance questions and claims. 800-390-3330.
- Missouri Division of Vocational Rehabilitation - assists individuals reasonable accommodations to remain in the workforce. 1-877-222-8963 or [www.vr.dese.mo.gov](http://www.vr.dese.mo.gov)



- Medicaid – health insurance administered through the state government. It is available to people over 65 and disabled persons who meet income and asset guidelines. Contact local Missouri Department of Social Services office for determination of eligibility.
- Missouri Statewide Independent Living Council -Advisory council appointed by the governor to monitor Independent Living Centers (ILC's) throughout Missouri. ILC's provide services to enable people with disabilities remain in their homes. The services vary between centers but may include assistance with home modifications, adaptive technology, equipment loan, advocacy, and information and referral. You can locate the ILC for your area at 1-877-222-8963 or [www.mosilc.org](http://www.mosilc.org).
- Statewide Recycling Computer Project – provides low cost, refurbished computer systems (CPU, monitor, keyboards, and mouse only) to person's with disabilities. Fees for program range \$60-\$300 based on applicant's household income. 800-647-8557 or 573-651-6464 or an application.
- Missouri Assistive Technology Project – provides loan equipment and low interest loans for augmentative communication needs. [www.dolir.state.mo.us/matp](http://www.dolir.state.mo.us/matp)

### **State of Missouri Reserved Parking Program**

A Disabled Person Placard (placard) is a removable windshield placard that is to be hung from the rearview mirror of a PARKED vehicle in order to park in disabled parking spaces. The placard must be used ONLY when a physically disabled person is the occupant of the motor vehicle at the time of parking or when the physically disabled person is being dropped off or picked up. The placard MUST NOT be hung from the rearview mirror when driving the vehicle.

Any physically disabled person, parent or guardian of a physically disabled person, a not-for-profit group or organization that transports more than one physically disabled person may be eligible.

The term "physically disabled" means a blind person, or a person with disabilities which limit or impair the ability to walk, as determined by a licensed physician as follows.

1. The person cannot walk 50 feet without stopping to rest;
2. The person cannot walk without the use of, or assistance from, a brace, cane, crutch, another person, prosthetic device, wheelchair, or other assistive device;
3. The person is restricted by lung disease to such an extent that the person's forced respiratory expiratory volume for one second, when measured by spirometry, is less than one liter, or the arterial oxygen tension is less than 60 mm/Hg on room air at rest;
4. The person uses portable oxygen;
5. The person has a cardiac condition to the extent that the person's functional limitations are classified in severity as Class III or Class IV according to the standards set by the American Heart Association; or
6. The person is severely limited in the ability to walk due to an arthritic, neurological, or orthopedic condition.

A physician, chiropractor, podiatrist, or optometrist must certify the person is permanently "physically disabled" by completing a Physician's Statement for Disabled Person's Placard

Complete an Application for Disabled Person Placard (DOR-2769) and have your physician complete a Physician's Statement for Disabled Person's Placard .These forms can be obtained at your local license office. Once the forms are completed, submit them along with the appropriate fee, either by mail to:

Driver and Vehicle Services Bureau  
PO Box 100,  
Jefferson City, MO 65105-0100

or in person at your local license office.

There is no fee required to obtain a Permanent Disabled Placard, and the placard expires on September 30th of the fourth year from which the placard was issued.

### **Transportation Issues Surrounding ALS**

Is my driving still safe and how will I get to where I need to go if I stop driving? -- are two of the hardest questions ALS patients ask themselves. Your personal safety and the safety of others should be your central concern. Technology and resources exist to make sure you have access to keep you mobile – but how do you know when it is time to let someone else drive? Most physicians will work with you and tell you to “use common sense”, but there are professionals that can help.

Professional driving assessment centers perform evaluations to help answer that question for you. Rehabilitation specialists recommend assistive devices (any tool that does a job for you) to keep you driving as long as possible. Assessments are extremely important to ensure personal safety, relieve stress of family members, help you save money, keep you insured, and overview any specialized concerns or needs you may have. Many private insurance companies cover driver rehabilitation programs. These professionals will work with your physician/occupational or physical therapist to elongate your independence.

Most communities offer handicap accessible (curb-to-curb) transportation; some car rentals offer handicap accessible vans for short-term rentals; private companies exist that will transport you (with/out wheelchair and extends to stretcher) to/from medical appointments. Another option is to modify an existing vehicle or purchase a handicap accessible vehicle. Some driver's insurance may cover vehicle modification <rarely> but the Veteran's Administration will help with the cost of adaptations. All vehicle manufacturers have a program to modify vehicles and most offer a rebate program when purchasing a new vehicle (up to \$1000). State Department of Vocational

Rehabilitation Services will assist with modifications while employed. Remember to check with an accountant regarding modification tax credits. **(this applies to not only your vehicle but home modifications too!)**

Almost any car can be modified, but the types of vehicles and modifications appropriate for each driver are based completely on individual need and preference.



**Always remember to obtain an electric wheelchair prior to modifying or purchasing an accessible vehicle.**

When choosing a vehicle for transportation, consider the following factors which may influence your decision: (Your doctor, physical therapist or occupational therapist will have input and can help answer any questions, but a certified adaptive driving specialist can assist in making a vehicle selection with completion of the driver rehabilitation program)

- Physical limitations - What will affect your ability to access a vehicle or drive? Can you handle driving stress?
- Ability to transfer into/out of a vehicle -Will you require an assistive seat or lift to get in or out? If you require a lift, what options do you prefer? Side door or rear door entrance/exit, electric/hydraulic, platform swingout or superarm, etc.?
- Special modifications - such as hand-controls, to operate the vehicle? Do you require a lowered floor or raised top and doors? What is your height? (head to ground sitting) and length/width of your wheelchair or scooter?
- Transportation needs - Do you need a car/truck/full-size or mini-van? How many will need to ride in vehicle?
- Services/warranty programs – parts covered? Where can repairs be done? Emergencies - if out of town?

More information is available at [www.unitedaccess.com](http://www.unitedaccess.com) or Contact your local ALSA chapter for a listing of resources/specialists in your area.

**Remember it is best to get a power wheelchair prior to getting a handicap accessible van as not all are compatible with each other!**

### **Private Insurance and ALS Benefits**

If you do not qualify for Medicare benefits or you choose to stay with your private health insurance, remember each insurance policy is individualized for patients and benefits will vary. Most Insurance providers follow the Medicare guidelines for services. However, it is important that you obtain a copy of your Certificate of Coverage. This document lists all services, benefits, and % of coverage for all components of your policy.

If you have been diagnosed with a fatal illness, including ALS, your insurance company will automatically assign a case manager to your file. It is important to obtain this person's name and contact number as it will speed up authorization for services. Also, with a fatal illness, the insurance company must review any appeals on denial of coverage within a 24 hour time period.

Remember, you can combine Medicare benefits (primary) and private insurance (secondary or supplemental) to get 100% coverage of services provided.

### **Long Term Care Insurance (LTC)**

Long-term care refers to a wide range of settings and services: health care, home care, adult day care, respite care, and various living facilities. If you have an LTC policy, it is recommended you read your certificate of coverage as well. This will answer questions regarding exact services and % of services covered. Most nursing home, skilled nursing facility, or custodial benefits can be redesigned

into home health services. This can be beneficial as most patients sometime throughout this disease process can benefit from additional help in the home.

As most ALS patients are cared for in their home by family members and friends, it is important to understand your LTC policy benefits. There are many services that can be provided while someone is residing in their own home. These programs encourage independence and offer support to the person with ALS and/or their family.

In-home options:

- chore services
- home maintenance or repair
- home modifications (grab bars, ramps, widened doors, etc.)
- medical equipment (walkers, scooters, bedside commode, etc.)
- meal services (Meals-on-Wheels)
- friendly visitors
- telephone reassurance
- respite care for the family care provider
- home health services
- adult day service programs
- personal attendants (for a few hours or full time)
- personal emergency response (like Lifeline)
- assistance from family, friends, neighbors (Share the Care Models)

If you do not have an LTC policy, many of these services are available in your community through religious groups, service and civic organizations, or local Area Agencies on Aging. Availability is dependent on your community resources. The ALS Association is also able to assist with some of these services.

If a skilled nursing facility is the best option for your long term care, remember to consult your ALSA case manager as special considerations apply for ALS patients living in a nursing facility. Your case manager can work with the staff to insure that all your needs are met.

**It may seem overwhelming as you move forward through the journey of ALS in trying to determine which resources and options are available to you. Remember that your ALSA case manager can help navigate through the maze and daze.**

### **Palliative Care Resources**

Supportive Care is an enhanced home care program developed for patients who are still exploring curative treatment options and need a stronger focus on pain management, family support, and spiritual care. This service combines home care services and palliative care professional and is often a preamble to Hospice with no time limit for this service. It can be a huge resource for patients with ALS.

Hospice is a special concept of care designed to provide comfort and support to patients and their families when a life-limiting illness no longer responds to cure-oriented treatments. Hospice care neither prolongs life nor hastens death. Hospice staff and volunteers offer a specialized knowledge of medical care, including pain management. The goal of hospice care is to improve the quality of a patient's last days by offering comfort and dignity. Hospice care is provided by a team-oriented group of specially trained professionals, volunteers and family members. Hospice addresses all symptoms of a disease, with a special emphasis on controlling a patient's pain and discomfort. Hospice deals with the emotional, social and spiritual impact of the disease on the patient and the patient's family and friends. Hospice offers a variety of bereavement and counseling services to families before and after a patient's death.

Before providing care, hospice staff meets with the patient's personal physician(s) and a hospice physician to discuss patient history, current physical symptoms and life expectancy. After an initial meeting with physicians, hospice staff meets with both the patient and their family. They discuss the hospice philosophy, available services and expectations. Prior to service, staff and patients also discuss pain and comfort levels, support systems, financial and insurance resources, medications and equipment needs. A "plan of care" is developed for the patient. This plan is regularly reviewed and revised according to patient condition. Bereavement services and counseling are typically available to loved ones for a year after the patient's death.

Most patients admitted to a hospice program are referred by their physicians. Others come themselves or are referred by the hospital, a community agency or by a friend or relative. Family members may request counseling from the hospice care program even if the patient does not wish to be admitted to the program. If you are interested in learning about or getting hospice care talk to your physician or contact a hospice provider in your area.

Hospice care is covered by Medicare and private insurance. Patients may be asked to meet co-pay or other uncovered costs. However, no one will ever be turned down for financial reasons. Hospice works in conjunction with the patient's physician providing care under a plan of treatment designed by the team in conjunction with the patient and family.

**Remember you can speak to a Hospice representative for a consultation before starting services.**

#### **Donation of Tissue for ALS Research**

*Why should you consider donating brain and spinal cord tissue for research?*



**Researchers have learned a great deal about amyotrophic lateral sclerosis (ALS), a fatal neuromuscular disorder, in the past dozen years. Yet, we still do not have all the answers as to what causes ALS or how it begins. Investigators are searching for the answers to these and many other puzzling questions about ALS in their effort to find effective treatments and ways to prevent the disease.**Because ALS is a disease of the motor neurons that are located in the brain and spinal cord, it is difficult to access this tissue in patients. Brain research using postmortem tissue is critical to understanding ALS. However, there can be a scarcity of ALS brain and spinal cord tissue for research studies.

Human tissue of ALS patients is the most precious resource for ALS researchers. ALS tissue when combined with complete information about the donor's ALS history will help investigators find answers. Researchers will compare ALS tissue with non-diseased tissue. They will compare tissue of patients with different histories - age of onset at time of diagnosis, progression of the disease, care and treatment during the course of the disease, and other vital information to unravel the mystery of why and how ALS begins and then progresses to a tragic end.

Persons with ALS and their families realize the importance of research into the cause of ALS, as understanding the cause offers the best opportunity for finding and producing effective therapies to treat and/or prevent ALS in the future. Many people have already played a key role in advancing the knowledge about ALS by providing blood samples, muscle biopsies and other specimens.

For some people, this decision or commitment to donate tissue upon their death may be difficult, especially when coping with the day-to-day challenges of ALS. However, it may offer a sense of purpose or comfort to know that one will be contributing to the continuing quest for knowledge about ALS.

Patients choosing to make this very generous donation need to plan ahead since there are a scarcity of centers prepared to perform the tissue donation procedures correctly. There is also a limited window of opportunity after death to realistically harvest tissue. Specific plans need to be made well in advance. If you are considering tissue donation, contact your physician at one of the centers listed below. Be sure to ask the center if they need ALS tissue at this time and can they store donated tissues. Also, be sure to ask if there will be any cost to you or your family. Hospice professionals can help your family make these arrangements.

### **ALS Research Centers that Utilize Donated Tissue**

Some of the ALS research centers geographically close that accept ALS tissue are listed below. Please contact these centers early in your planning process to ascertain if they can utilize your donation.

#### **Illinois**

Northwestern University, Department of Neurology  
Medical School Tarry 13-715  
303 E. Chicago Ave., Chicago, IL 60611-3008  
Nailah Siddique, RN  
Nsiddique@northwestern.edu  
312-503-2712

#### **Kentucky**

University of Kentucky, Department of Neurology  
Kentucky Clinic, L445, Wing D, Lexington, KY 40536-0284  
Edward J. Kasarskis, MD, PhD, Director  
859-281-4920

**Resources for organ and tissue donation other than brain and spinal cord tissue are:**

**The National Disease Research Interchange**

1880 JFK Blvd., 11th Floor, Philadelphia, PA 19103  
Lisa Morris, Sr. Mgr. Odyssey Dept.  
800-222-6374 ext. 245

**Gift of Hope (IL/IN)**

660 N. Industrial Dr  
Elmhurst, IL 60126  
630-758-2600

They will direct the donation to anywhere in the country.

**Center for Organ Recovery and Education (CORE)**

204 Sigma Dr., Pittsburgh, PA 15238  
Pat Kornick, Director of Awareness and Education  
800-DONORS-7 \*

CORE's National office staff will direct the patient and family to a procurement center in their geographic area. Be aware that in many cases, organ and tissue donation - except for brain and spinal cord - may not be possible for people with ALS as patients must be declared "brain dead" first. Consult CORE, your local organ and tissue center, or your neurologist for specific and individual information.

**General Research Donation**

Another avenue of donating tissues, organs, or body to science for research is to a local medical school. Tissue and bodies are used for general research purposes and training of surgeons, with no guarantee that it will aid in ALS advancements. These arrangements must be made prior to time of death. Specific requirements are made with each receiving institution. Hospice professionals can make arrangements with your family.

We have attempted to highlight the main issues that surround ALS in this informational manual. For all other questions, please contact the chapter office as a case manager is available to answer questions, provide direction, and meet with your family to help you live longer and stronger during your course of ALS.

Also, please visit [www.alsa-stl.org](http://www.alsa-stl.org) for other life management tips and resources located on our chapter website.

**Please complete the following form and return to the office in the envelope provided to enroll for services through The ALS Association St. Louis Regional Chapter. All information is kept confidential as the chapter follows the guidelines of The Health Insurance Portability and Accountability Act**

- If you need assistance in completing this form, please call the chapter office and a Care Services and Outreach Specialist can meet with you to accommodate this need.

**This form is also available online at [www.alsa-stl.org](http://www.alsa-stl.org)**

# Patient Intake Information

Last Name: \_\_\_\_\_ First Name: \_\_\_\_\_ Middle: \_\_\_\_\_  
Street: \_\_\_\_\_ City: \_\_\_\_\_ State: \_\_\_\_\_  
County: \_\_\_\_\_ Zip + 4: \_\_\_\_\_  
Home Phone: \_\_\_\_\_ Cell Phone: \_\_\_\_\_  
Email: \_\_\_\_\_  
Date of Birth: \_\_\_\_\_ Race: \_\_\_\_\_  
Are you registered with the National ALS Registry at [www.alsa.org/als-care/als-registry/](http://www.alsa.org/als-care/als-registry/)? \_\_\_\_ Yes \_\_\_\_ No

Marital Status: \_\_\_\_ Single \_\_\_\_ Married \_\_\_\_ Separated \_\_\_\_ Divorced  
\_\_\_\_ Wid. \_\_\_\_ Other

## **PRIMARY CAREGIVER**

Name \_\_\_\_\_ Relationship \_\_\_\_\_  
Email \_\_\_\_\_  
Address if different: \_\_\_\_\_  
Date of Birth \_\_\_\_\_ Home Phone \_\_\_\_\_ Cell Phone: \_\_\_\_\_

## **OTHERS LIVING IN HOUSEHOLD:** (Please use additional sheet of paper if necessary)

1. Name

\_\_\_\_\_

Date of Birth	Race	Email Address
_____	_____	_____

2. Name

\_\_\_\_\_

Date of Birth	Race	Email Address
_____	_____	_____

## **EMPLOYMENT INFORMATION**

Are you presently employed? \_\_\_\_ Yes \_\_\_\_ No \_\_\_\_ Retired  
Retirement Date \_\_\_\_\_  
Most Recent Employer and Position: \_\_\_\_\_

Are you a veteran? \_\_\_\_ Yes \_\_\_\_ No What branch of Service \_\_\_\_\_  
Service Dates \_\_\_\_\_  
How did you hear about us? \_\_\_\_ Hospice \_\_\_\_ PCP \_\_\_\_ Neurologist \_\_\_\_ MDA  
\_\_\_\_ United Way \_\_\_\_ Website  
\_\_\_\_ Other Please  
list: \_\_\_\_\_  
\_\_\_\_\_

**MEDICAL INFORMATION**

Date of Diagnosis \_\_\_\_\_ Is there any other person(s) in your family with

ALS? \_\_\_\_ Yes \_\_\_\_ No

If so, what is their relationship to you? \_\_\_\_\_

Neurologist: \_\_\_\_\_ Address \_\_\_\_\_

Phone \_\_\_\_\_

Primary Physician: \_\_\_\_\_ Address \_\_\_\_\_

Phone \_\_\_\_\_

**OTHER CHILDREN/SOCIAL SUPPORT:** (Please use additional sheet of paper if necessary)

1. Name \_\_\_\_\_

Address \_\_\_\_\_ Date of Birth \_\_\_\_\_

\_\_\_\_\_

Phone \_\_\_\_\_ Email Address \_\_\_\_\_ Race \_\_\_\_\_

\_\_\_\_\_

2. Name \_\_\_\_\_

\_\_\_\_\_

—

Address \_\_\_\_\_ Date of Birth \_\_\_\_\_

\_\_\_\_\_

Phone \_\_\_\_\_ Email Address \_\_\_\_\_ Race \_\_\_\_\_

\_\_\_\_\_

**ASSISTIVE DEVICES CURRENTLY IN USE**

\_\_\_ Bedside Commode \_\_\_ Communication Device \_\_\_ Hoyer Lift \_\_\_ Manual

Wheelchair \_\_\_ Tub Seat

\_\_\_ Cane \_\_\_ Hospital Bed \_\_\_ Lift Chair \_\_\_

Powered Wheelchair \_\_\_ Walker

Other (describe): \_\_\_\_\_

**OTHER INFORMATION:** This information is gathered for United Way statistical reporting purposes only. It does not affect your ability to receive chapter services.

What is your average household income? Please check one.

\_\_\_ \$0-\$9,999 \_\_\_ \$15,000-\$19,999 \_\_\_ \$30,000-\$49,999 \_\_\_ \$100,000 and greater

\_\_\_ \$10,000-\$14,999 \_\_\_ \$20,000-\$29,999 \_\_\_ \$50,000-\$99,999 \_\_\_ Unknown

## ***Local Evaluation Facilities for Augmentative Communication Devices***

**Make sure your provider of choice is approved by your insurance/Medicare/Medicaid**

St. Johns Hospital  
Heidi Hockstetler  
800 E. Carpenter  
Springfield, IL 62761  
217-544-6464 ext. 44871

Barnes Jewish Rehabilitation  
Miranda Weiss, CCC - SLP  
#1 Barnes Jewish Hospital Plaza  
St. Louis, MO 63110  
314-362-2381

St. Elizabeth Hospital  
Voice Clinic  
Susan Carpenter  
211 S. 3<sup>rd</sup>  
Belleville, IL 62220  
618-234-2120 ext. 1235

Missouri Rehabilitation  
Assistive Technology  
600 N. Main  
Mt. Vernon, MO 65712  
800-466-3711  
417-461-5354

Missouri State University  
Communication Sciences/Disorder  
901 S. National Ave.  
Springfield, MO 65810  
417-889-6223

Laura Chandler, CCC\_SLP  
Memorial Hospital of Carbondale  
405 West Jackson St. Carbondale, IL 62901  
618-549-0721

Fontbonne University  
Dept. of Communication Disorders  
Gail Rice, CCC/SLP  
6800 Wydown Blvd.  
St. Louis, MO 63105  
314-889-1407

Eastern IL University  
Dept. of Communication Disorders  
600 Lincoln Ave.  
Charleston, IL 61920  
217-581-5000

Assistive Technology Assessment Center  
Michelle Wheeler, SLP, Shawna Dunnaway, SLP  
201 Business Loop 70 West, AP Green Bldg, Room 125  
Columbia, MO 65203  
573-882-9111  
in home assessments available

**For additional resources please see the Assistive Technology Manual**

