

LIVING WITH ALS

Managing Your Symptoms and Treatment



Cover: *Maze* by Mary Louise Hartnagel.
The artwork on the covers of the *Living With ALS*
Manuals was created by individuals with ALS or
their family members.

About the Artist

“Mary Louise Hartnagel, my only sister, died in 1994 from the bulbar form of ALS. She was 47 years old, single with no children.

Art was everything to Mary. She had an Associate’s Degree in Art and could see beauty, artistic beauty, not only in known works of art, but also in everyday life. She often studied other artists by replicating their work. Her “works of others” were greater than the art she created herself, yet she displayed them all tastefully in her home in different “rooms”: A Flower Room, Native American Room, Bird Room, etc. Each room contained artwork and items of all media. She could find beauty in many things: chipped vases, old wood moldings, furniture pieces, old houses, clothing and hats. She wore a tasteful, sometimes flamboyant hat everywhere she went.

I deeply thank The ALS Association for honoring Mary’s memory by sharing her work and her intense personal love of art with others who may be facing the same path in life that she did.”

ROBIN HARTNAGEL
*Loving sister and caregiver of
Mary Louise Hartnagel*

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Manual 3

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Written by

Deborah Gelinas, MD

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

LIVING WITH ALS

**Managing Your Symptoms
and Treatment**

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Contributors to the 2002 edition:

M. L. Del Bene, MS, RN, NP-P
Deborah Gelinas, MD
Hiroshi Mitsumoto, MD
Susan Walsh, RN, MSN

Project Advisor:

Mary Lyon, RN, MN

Graphic Design:

Denton Design Associates

Illustration (text pages):

Neverne Covington

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Introduction

W

hen you were first given the diagnosis of ALS, you may have felt overwhelmed and powerless. Your life suddenly seemed out of control, but it does not have to stay that way. You can be in charge of your life again, and to some extent gain control over ALS.

Many of the symptoms you suffer from can be treated effectively. By learning more about ALS and your symptoms, you can become empowered. The weakness due to ALS must be accepted, but you do not have to put up with symptoms like depression, insomnia, and muscle cramping which can be eased with medication and other helpful methods.

The greatest challenge of living with ALS is how to live with it day to day. This manual will review many of the challenges you may face. It offers a comprehensive look at the symptoms you might experience and explains the treatment options available, which can help improve the quality of your life. Not all of the symptoms mentioned here are experienced by every person with ALS. Take what applies and leave the rest. If some symptoms do sound familiar to you, read more about them and share the suggestions offered with your caregivers, doctors, nurses, therapists, friends, and family.

What Are the Possible Causes of ALS?

A

myotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease which results in the death of motor nerve cells in both the brain and spinal cord. This loss may lead to paralysis of all muscles under voluntary control, including those for your face, mouth, arms, legs, back, and breathing.

You have been diagnosed with ALS because of specific symptoms which have worsened over time, such as:

- muscle weakness, wasting, cramping, and/or twitching
- slurred, slow speech
- difficulty in swallowing
- inability to control emotions
- slow and uncoordinated movements

There is no one specific test to confirm the diagnosis. After observing your symptoms over a period of time and ruling out other possible diseases, both you and your doctor have become convinced that you have ALS.

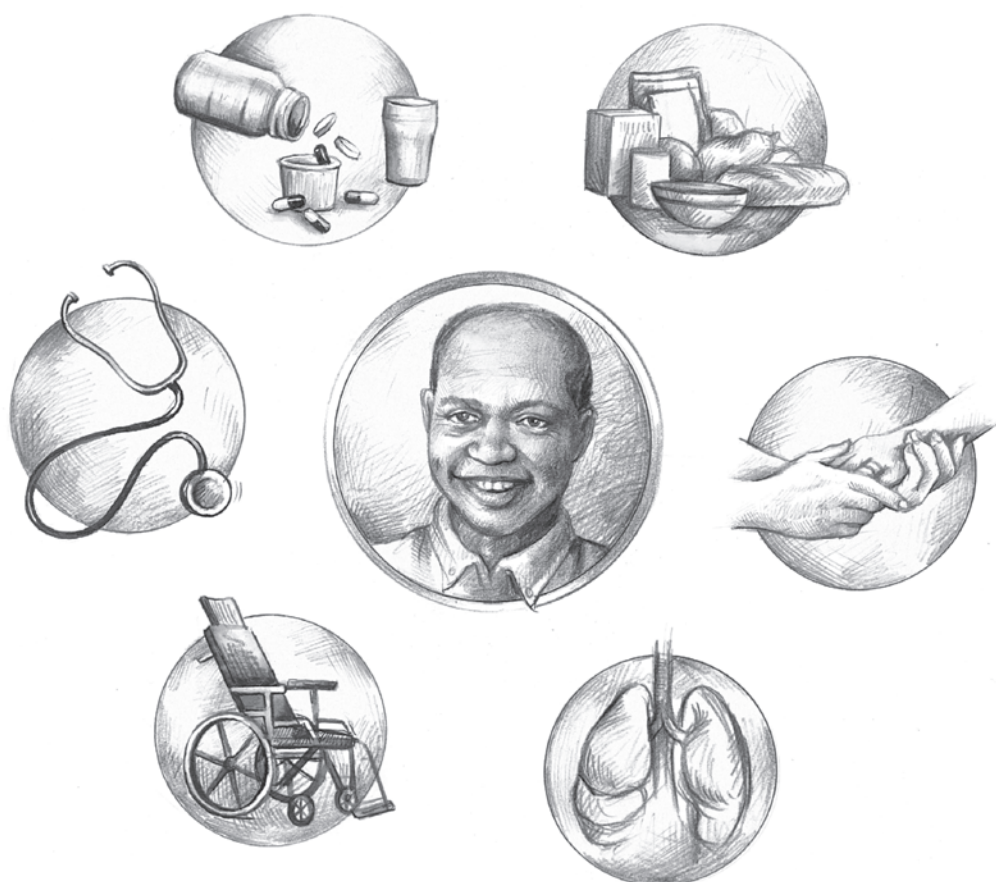
The cause of ALS remains unknown although much more about what goes wrong in the body has been learned over the past decade. Many different theories have been considered, and a great deal of exciting research has been completed and continues to be undertaken, which begins to shed light on the mystery of this disease.

It is becoming clearer that ALS is likely a disease with more than one cause. Many factors are known or theorized to play important roles in its ultimate cause and progression.

Despite the explosion of new scientific findings, the root cause or causes for ALS are not yet known. However, research is focusing on the following areas.

- Excitotoxicity (such as that caused by excess glutamate)
- Inflammation/Inflammatory Response
- Oxidation/Oxidative Stress
- Protein Aggregation (abnormal accumulation of proteins)
- Axonal Transport Abnormalities
- Neurofilaments
- Mitochondria
- Environment
- Risk/Susceptibility Genes
- Apoptosis (premature programmed cell death of the motor neurons)

Some of the above areas may be factors that only come into play and promote progression of the disease after the initial trigger – like a domino effect. (See Manual 1 for more information.)



Living with ALS requires an all-encompassing program of treatment and support.

What Are Your Symptoms and Treatment Options?

P

PRACTICE GUIDELINES FOR ALS

The Quality Standards Subcommittee of the American Academy of Neurology undertook an extensive review of the medical literature to determine recommendations for the clinical management of ALS based on peer-review publications. The practice parameters for ALS were developed to improve the care and the quality of life of people with ALS by providing a rational basis for managing the disease.

Five clinical areas were investigated:

1. Informing patients and families about the diagnosis and prognosis (Breaking the news)
2. Symptomatic treatment
3. Nutrition and decisions about a feeding tube
4. Respiratory problems and decisions about non-invasive and invasive (tracheostomy) ventilation
5. Advance directives and palliative care

In each of these five clinical areas, specific recommendations are provided for the clinical management of ALS. A summary of these recommendations can be found in the table below.

It is important to discuss your individual care and the practice parameter recommendations with your physician and health care team. Learn what the treatment alternatives are and what options are available for the symptoms you are experiencing. Ultimately, each decision about treatment interventions is yours to make.

Recommendations for Clinical Management of ALS

Summarized from: Practice Parameter: The care of the patient with amyotrophic lateral sclerosis (an evidence based review), *Neurology* 1999; 52:1311–1323.

Breaking the News

- The physician should give the diagnosis to the patient and discuss its implications. The patient's social and cultural background should be respected in the communication process.
- The diagnosis should always be presented in person.
- Printed materials about the disease should be provided and the patient told about support and advocacy organizations.
- Physicians should present the diagnosis in a timely manner, provide sufficient information, deliver the news in a caring and supportive manner and provide hope.

Excessive Saliva

- Treat with one of the several medications known to be effective.
- Treat thick mucus with one of the known effective medications.
- Consider offering manually assisted coughing and mechanical insufflation for helping patients clear secretions from the throat and lungs.

Emotional Lability (exaggerated crying or laughing)

- Treat with one of the known effective medications.

Nutrition

- PEG feeding tube is indicated for people with ALS who have significant difficulty swallowing and experiencing weight loss.
- For optimal patient safety as well as effective nutrition and hydration, generally PEG should be offered while the patient's vital capacity is more than 50% of the predicted value.

Breathing Problems

- Physicians should monitor the patient for symptoms of reduced breathing volume (hypoventilation).
- Offer noninvasive ventilatory support as an effective initial therapy for symptomatic chronic hypoventilation and to prolong survival in patients with ALS.
- When longer survival is the goal, offer invasive ventilation and fully inform patient and family of burdens and benefits.

Palliative Care

- Physicians should respect the right of the patient with ALS to refuse or withdraw any treatment, including mechanical ventilation.
- When withdrawing ventilation, provide medications to relieve any difficulty breathing and anxiety.
- Provide treatment for reversible causes of difficulty breathing, if present.
- Provide medications for non-reversible difficulty breathing to reduce suffering.
- Provide medications to alleviate pain, if present.
- Consider referral to hospice.
- Discuss advance directives well in advance of the terminal phase and reevaluate with the patients every six months.

For more printed information about the ALS practice parameters, you and your physician can contact The ALS Association at (800) 782-4747.



The goal of the ALS Clinical Assessment, Research and Education (ALS C.A.R.E.) program is to serve as a resource for measuring and improving care for patients with ALS and their caregivers. The program also develops educational projects and conducts research.

An integral part of the ALS CARE program is the ALS Patient Care Database, a voluntary, confidential, North American database of clinical outcomes of patients with ALS and their caregivers. The database provides a mechanism for neurologists to evaluate how their patients respond to various clinical treatments. By collecting accurate and timely data on patients and their caregivers, participants make a significant contribution to developing the ALS knowledge base in the areas of making the diagnosis, predicting survival, risk factors for the disease, functional status, quality of life, treatments and the management of symptoms.

To learn more about the ALS CARE Program and findings-to-date, visit the web site at <http://www.unassmed.edu/outcomes/als/>. If you are interested in participating in the ALS CARE Database, discuss this with your physician.

Cramps/Spasms

A muscle cramp is a sudden **involuntary muscle contraction** (not under your control) that may be triggered by voluntary exertion of any muscle group in your arms, legs, chest, back, abdomen, jaw, or throat. Cramps are caused by a brief contraction of a weakened muscle due to an explosive over-activity of motor nerves.

These contractions have a range of severity including mild, intermittent and severe. They may be extremely painful, prolonged, and interfere with waking activities, nighttime sleep, or they can occur mild and intermittent. When having a cramp, you may notice visible knotting in your muscle and abnormal posture until it passes. You can reduce the number of these cramps and relieve them by doing stretching exercises and by staying well-hydrated with mineral-balanced drinks such as Gatorade®.

If your cramps are very severe, you should talk with your doctor about specific prescription medications available to help relieve the symptoms. Some medications that may be recommended by your doctor include baclofen, clonazepam, lorazepam and gabapentin. New medications for the treatment of muscle cramps are being studied.

Fatigue

You may be aware of general muscle fatigue and exhaustion since they are common features in ALS. The cause may be the extra burden placed upon your surviving nerve cells. As neurons die, remaining ones send signals to activate the otherwise unused muscle, and a single surviving neuron may be doing 100 times its normal workload.

Thus, there may be times when you just cannot do a certain task (such as climbing stairs), which when rested you are able to perform with very little difficulty. You may be exhausted by the end of the day, thereby having to pace yourself and take naps. If you do very heavy activities on one day, you might find that you will “pay for it” the next day.

Although medications may be prescribed for fatigue they are seldom effective. The best treatment for fatigue is proper pacing of your activities and conservation of your strength. Ask your doctor about medications which may provide temporary relief for fatigue, such as Mestinon[®], Ritalin[®], Symmetrel[®], and Cylert[®].

Loss of Control Over Emotions (Pseudo-Bulbar Affect)

Perhaps you now have difficulty controlling your emotions, crying easily or laughing at inappropriate times. This reaction is called emotional lability and can include excessive crying or laughing beyond what the person is actually feeling. It is thought to be caused by the lack of restraint of the bulbar area of the brain involved in laughing and crying. You may want to discuss with your doctor the various medication options such as amitriptyline, serotonin, reuptake inhibitors such as paroxetine, fluoxetine or fluvoxamine with your doctor. A promising new treatment, Neurodex (Avanir Pharmaceuticals), is awaiting approval by the Food and Drug administration following clinical trials. An open-label phase is ongoing as of the date of publication. While it remains an investigational drug, results from an earlier Phase II/III clinical trial that included 140 persons with ALS, showed

Neurodex decreased the severity and frequency of pseudobulbar episodes and improved quality of life.

Urinary Urgency

You may find that you are having difficulty holding in your urine and “making it to the bathroom on time.” This problem can be due to many reasons: Your brain may be giving you the “signal” too late that your bladder is full; you may be taking longer to reach the bathroom and to arrange your clothing; or you even may have a bladder infection (unrelated to your ALS).

Discuss this symptom with your doctor, because he/she may want to check for a bladder infection. If you do not have an infection, then your doctor may prescribe medication to relieve your symptoms, such as oxybutynin or tolterodine, which can help you hold your urine longer.

Swelling of Hands and Feet

Normally, blood is returned to the heart through the pumping action of muscles, but a paralyzed or weakened limb is not able to effectively pump back the blood. This problem may cause your feet to become puffy (especially if you tend to be sitting longer because of leg weakness), or your fingers may swell due to hand weakness. To reduce swelling, elevate the affected limb above the level of your heart whenever possible. You also may find that support stockings are helpful, for both men and women.

If swelling is painful, or fails to decrease after you have tried elevating your arm or leg, please discuss this condition with your doctor. Occasionally, a swollen limb may indicate the presence of a blood clot, which if left untreated can dislodge and travel to your lungs; the result is a **pulmonary embolus** which causes shortness of breath and can be life-threatening. Once this condition is identified, it can be treated effectively by taking the proper medication.

Deep Vein Thrombosis

A deep vein thrombosis (DVT) is a blood clot that usually develops in the leg and occurs when the blood flow slows down or the vein is damaged. DVT can be painful and cause further harm to a person's lungs if not treated. To reduce pain, people are encouraged to elevate their feet and legs by placing them on a pillow or cushion. Compression stockings, which are tighter than the average sock, are also offered as a form of treatment for pain and must be worn for several months. DVT may occur in people over the age of 40, or who are obese, or immobile or paralyzed. An anticoagulant is most commonly given to people with a DVT to help thin the blood. These are taken for three to six months. These medications do not reduce existing clots but can help new clots from forming and existing clots from growing.

Many people living with ALS, who are immobile or sit for long periods of time may want to look for signs of clotting in order to prevent them. Also, if traveling long distances, it is a good idea to discuss the possibility of DVT prevention with your health care provider.

Drooling/Salivation

You might experience pooling of saliva in your mouth, which can make you more likely to drool, swallow secretions, or cough. It may seem as though your body is producing too much saliva, but it is not. The body normally produces large quantities in the course of a day, helping to maintain a healthy mouth and good digestion. For reasons that are not clearly understood, you may not be swallowing your saliva as automatically (without thinking) as you did prior to ALS. If you have difficulty swallowing, saliva can collect in your mouth and cause drooling. In addition, you may have even more saliva when you are hungry, anxious, or smelling good food.

If your saliva is bothersome, there are several medications prescribed to help dry out your mouth, including glycopyrrolate, amitriptyline, diphenhydramine and hyoscyamine.

Thick Phlegm/Postnasal Drip

You may be breathing through your mouth more often than before, especially if your nose is congested. Mouth-breathing causes saliva to dry out and thicken (**phlegm**), and the medications taken to reduce drooling can cause excessive dryness and concentrated secretions. If phlegm becomes thick and sticky, you may hear a “gurgling” in your voice or have a feeling of “frog in your throat.”

If you are taking medication to reduce your saliva, decreasing the dose (after consulting with your doctor) may be helpful, as may adding a room humidifier or increased fluids. Other medications which your doctor may prescribe to clear your throat are guaifenesin (tablets), Organidin® liquid, or Robbitusin® (OTC) liquid. Inhalation breathing treatments also may help; a device called a **nebulizer** or an IPV (Intrapulmonary Percussive Ventilator) machine can be used to administer inhaled medicines. Another very helpful device is an ABI vest, which can be used for 10-minute intervals to help bring up secretions.

When allergies contribute to **postnasal drip** (secretions behind the nose), medications such as diphenhydramine or over the counter antihistamines in addition to nasal inhaled steroids may also be helpful.

Jaw Quivering

Your jaw may have a tendency to quiver or your teeth to chatter when you are chilled, yawning, or even speaking. If this condition becomes bothersome, some helpful medications your doctor may recommend are lorazepam or clonazepam.

Laryngospasm

Tightening of muscles in the throat (**laryngospasm**) with closure of the vocal cords and a sudden gasping for breath can be a frightening symptom. For a brief time, you may feel as if your airway is cut off and you will suffocate. Factors which may trigger laryngospasm include smoke, strong smells, strong alcohol, cold or rapid bursts of air, spicy foods, liquids or saliva swallowed “down the wrong pipe,” and stomach acid **regurgitation** (backward flow to the mouth).

If this type of reaction does occur, you should remember that laryngospasm will pass on its own. You can achieve immediate relief by dropping your chin down to your chest and swallowing, by breathing slowly through your nose, and by opening a window or door to provide fresh air.

You may take antacids to reduce the contribution of gastric reflux (heartburn) which may trigger these attacks of laryngospasm. Such medications (both over-the-counter and prescribed) include antacids such as omeprazole, ranitidine, famotidine and a variety of newer drugs. If laryngospasms persist despite careful avoidance of identified triggers, ask your doctor about prescribing lorazepam to lessen the spasm.

Acid Indigestion

You do not need to have a diagnosis of ALS to suffer from acid indigestion and heartburn, but if you have these symptoms *and* ALS, you may be more likely to experience **gastro-esophageal reflux disease (GERD)**. GERD can be masked by a cough, bad breath, throat irritation, voice hoarseness, shortness of breath, nausea, and insomnia. Since these particular symptoms of GERD can occur without heartburn, you may not realize that you have this problem.

Heartburn is caused by stomach acids flowing back up (reflux) into the esophagus. Normally, these stomach acids are prevented from entering the esophagus by the strong muscles in your diaphragm. Caffeine, spicy foods, overeating, and diaphragm weakness all increase acid reflux into the esophagus.

Some people experience heartburn more significantly after being nourished through a feeding tube, because they are suddenly being overfed after weeks of eating very little by mouth. Speak to your doctor about treating GERD with medications such as metoclopramide.

Nasal Congestion/Ear Blocking

If there is bulbar (speaking, swallowing, upper airway, and clearing of saliva) involvement from ALS, you may notice that your nostrils become plugged up, especially at night. This condition is due to weakness of the muscles which normally elevate the nostrils and open the airways. Likewise, these muscles may fail to maintain tone in your **eustachian tubes**, which connect the mouth and ear canals, giving rise to a feeling of blocked ears.

Elevating your nasal bridge with Breathe Right® tape used by athletes can relieve your discomfort significantly. Also, avoid wearing heavy eyeglasses which depress the nasal bridge/cavity. Over-the-counter antihistamines/decongestants, Afrin® and Flonase® nasal spray can provide relief.

Constipation

Constipation can be a most uncomfortable symptom in ALS, resulting in hours spent upon the commode, abdominal pain, nausea, and distress. The causes include: decreased fluid, fruit, and vegetable consumption in your diet; less exercise; and reduced ability to bear down with the abdominal muscles. Once constipation starts, it can be a vicious circle until a good bowel regimen is restored. You may require stool softeners, laxatives or Fleet® Enema (as needed), options that are far preferable to hospitalization for intestinal obstruction and **bowel impaction** (overloading of stool/feces).

A careful review of medications taken to control saliva or pain may reveal that some of your constipation is due to their use. Discuss this possibility with your physician.

Careful management of all factors contributing to constipation in ALS, such as adequate water and fiber, stool softeners and laxatives, and an increase in activity or range of motion exercises can result in a bowel program that works for you.

“Power pudding” can be a helpful recipe for constipation. It consists of equal parts of prunes, prune juice, applesauce, and bran. Two tablespoons with each meal and at bedtime should be taken as a preventive measure along with increased fluid. For acute constipation you can try Mylanta®, over-the-counter laxatives, rectal suppositories, or a Fleet® Enema. Manual disimpaction may be required, but with a good bowel maintenance program and toilet aids (raised toilet seat / bedside commode) this method rarely becomes necessary.

Sleep Disturbances

Many factors may contribute to **insomnia** (disorders of initiating or maintaining sleep) and frequent awakenings. If you suffer from sleep disturbances, it is extremely important to understand the cause, not just to disguise the symptoms with medications.

The best method to understand your insomnia is to undergo a sleep study. If this procedure is not practical, a **nocturnal oximetry** should be obtained, which will monitor your heart rate and oxygen saturation throughout the night by a simple taped-on finger electrode. In this way, more serious sleep disturbances such as **central** and **obstructive apnea** (conditions where breathing stops during sleep) may be excluded from the possible causes.

Central apnea specifically refers to the change in breathing patterns that may occur due to alterations in the brain during sleep, especially dream sleep; it is best treated by noninvasive ventilatory support, such as positive pressure (NIPPV). NIPPV uses a machine connected through nasal tubes or a mask that provides pressure to the upper airway to keep it open and stimulate deep breathing.

Obstructive apnea refers to a blocked upper airway caused by nasal congestion, excessive tongue and jaw relaxation, or intermittent laryngospasm. These problems are more common if you have bulbar ALS, causing impaired motor nerve function that may make the upper airway close during sleep. This problem can be worsened by sedatives or alcohol.

Collapse of the upper airway will cause you to awaken frequently throughout the night, feel groggy in the morning and/or sleepy throughout the day, and also have trouble concentrating.

Obstructive apnea symptoms are also well-managed by NIPPV which applies pressure inside the nose and mouth to keep the upper airway open and allow freer breathing. Other simple measures that help with these symptoms are elevating the head of the bed, sleeping in a comfortable recliner, or lying on your side instead of flat on your back.

Another factor leading to insomnia may be periodic leg movements—spasms or kicks of the leg which are sufficient enough to awaken you (as well as your bed partner) throughout the night. It may be treated by such medications as carbidopa, levodopa, clonazepam and codeine.

Anxiety and depression also may cause insomnia, especially common shortly after diagnosis. If your sleep is continually disturbed, ask your doctor to prescribe a mild relaxant or anti-depressant such as Elavil®, Zoloft®, or Desyrel®, Buspar®, amitriptyline, fluvoxamine or desipramine.

If you have advanced ALS and sleep and comfort are main concerns, discuss with your doctor using the following at bedtime: Zolpidem, temazepam or even oral morphine.

Depression/Anxiety

It is entirely understandable that you may feel depressed after receiving a diagnosis of ALS. Some of the symptoms include daily sadness, loss of interest in friends and hobbies, irritability and anger, and sleep and appetite problems. Discuss your symptoms with your doctor and/or ask for a referral to a therapist so that you receive appropriate treatment. A psychiatrist, psychologist, clinical social worker, psychiatric nurse, or a therapist can help you to cope with the problems and emotional issues you might experience.

A one-month trial of an antidepressant may be appropriate. While it will not change the reality of ALS, the medication will decrease your feelings of helplessness and hopelessness and make your diagnosis seem more bearable. Your doctor can assist you in choosing the best antidepressant for you, such as fluoxetine, fluvoxamine, paroxetine or bupropion.

If depression leads to your contemplating suicide, an antidepressant is especially recommended along with supportive psychological counseling. After several weeks on the medication, you should begin to feel significantly more hopeful.

Mental Status

Although most people with ALS do not develop any cognitive changes, new research is demonstrating a link between ALS and a spectrum of frontotemporal lobar dementia (FTLD). These cognitive changes are occurring more frequently than previously believed and range from mild to moderate change. Cognitive changes are inclusive of, but not limited to, the inability to focus, lack of insight or poor judgment and mood or personality changes. Other symptoms may include difficulty with word generation, memory or other executive functions. Once more is known about FTLD, interventions and strategies may be developed to help patients, family members and caregivers to lessen the severity of the episodes and continue to have a prolonged quality of life.

Swallowing Difficulties

While this topic is discussed at length in Manual 5, it merits three recommendations:

- Tuck your chin down while swallowing.
- Swallow two-to-three times for every mouthful of food.
- Avoid foods which cause you the greatest difficulty swallowing.

Speech Problems

These concerns also will be addressed in-depth in Manual 5, but three quick tips are:

- Speak slowly.
- Over-exaggerate like a “bad Shakespearean actor.”
- When stuck, spell out the word.

Shortness of Breath

This topic is covered extensively in Manual 6; however, if you have shortness of breath or other difficulties breathing, speak to your doctor concerning a trial of noninvasive positive pressure ventilation (NIPPV). These nasomechanical ventilators, help inflate the lungs when your breathing muscles become weak. NIPPV can be used from a few hours per day to constant use. It is not invasive and you can make the decision each day to use it or not.

If you are advanced in your disease and have made the decision that care and comfort are your sole concern, then your doctor may prescribe oxygen and medications such as lorazepam and oral morphine, as needed to relieve any “air hunger.”

What Are Your Specific Medication Options?

A

lthough there is currently no cure for ALS it is not uncommon to live for many years. In the course of the disease, you may develop symptoms other than those typically described. While there is no satisfactory treatment for the most prevalent symptom, *weakness*, many good therapies are available for some of the other common ailments.

RILUTEK® (RILUZOLE)

This glutamate-blocking drug developed and manufactured by Aventis is the *very first drug approved* (December 12, 1995) exclusively for the treatment of ALS by the Food and Drug Administration (**FDA**). Rilutek® has been proven effective in well-controlled studies involving more than 1,100 ALS patients worldwide.

Preclinical data suggest that Rilutek® protects motor neurons from degeneration and death. The patients who received this drug at 50 milligram twice a day were shown to have a modest prolonged survival time compared to patients who were given a **placebo** (an inactive substance). Specifically, the clinical evidence demonstrated that this medication extends survival of **tracheostomy-free** (no invasive breathing support) ALS patients.

Generally, Rilutek® is well tolerated; however, its most common side effects are fatigue and upset stomach. If your doctor prescribes Rilutek®, be sure to start slowly, and take it orally with food so that you reduce stomach reactions. Two initial clini-

cal trials showed that Rilutek treatment is associated with modest increases in patient survival. A third clinical trial included older and more severely affected patients and did not show the same survival benefits as the first two clinical trials.

Recent review of data from two ALS clinic databases (U.S. and Ireland) suggests that Rilutek use in ALS may be associated with longer survival beyond the modest improvement shown in the clinical trials. More research is needed.

For each of the first three months on Rilutek[®], you must have your doctor check your blood (do a complete blood count) and liver functions, and thereafter every three months. This care is recommended because Rilutek[®] sometimes can cause changes in blood count or liver functions. For any further information or answers to specific questions about Rilutek[®], call 1(800) 790-RTEC (7832).

STATUS OF OTHER TREATMENT DEVELOPMENTS

Myotrophin[®] or IGF-1

This nerve growth factor, produced by Cephalon, also has been evaluated in two large clinical trials of ALS patients. The first, in North America, reported that patients receiving Myotrophin[®] had less ALS disease progression and a better quality of life than did patients receiving a placebo. The second trial, completed in Europe, yielded negative results. A third clinical study of Myotrophin[®] is underway. Myotrophin is not approved yet by the U.S. FDA for treatment of ALS.

This drug is injected **subcutaneously** (beneath the skin, like allergy or insulin injections) twice a day, and the most frequently reported side effects are redness, pain, and swelling at the injection site.

Creatine

Creatine is a naturally occurring biochemical involved in the energy metabolism of muscle. Past laboratory studies suggested that oral creatine supplementation may benefit patients with ALS. One U.S. multi-site clinical trial of creatine is completed showing no benefit for ALS. Another study is in process and the first results are expected in late 2005. One European study of creatine reported no benefit for people with ALS.

Minocycline

Caspase enzymes are thought to play a role in cell death in ALS. Minocycline is known to inhibit caspase enzyme activity. Minocycline has been shown to slow deterioration in animal studies of neurodegenerative disorders with cell death mechanisms similar to those in ALS. A safety and efficacy trial (in humans) as a treatment for ALS was completed and a larger multi-site trial began in 2003 and is ongoing.

Celebrex

Studies of this drug (approved by the FDA for the treatment of arthritis) in mouse models of ALS have shown neuroprotective effects and prolonged survival. There was a multi-center study of Celebrex in ALS patients, and the initial analysis of data from a 12 month study failed to show a benefit.

For more information about treatment development for ALS, visit The ALS Association's Drug Development Update in the Research Section of our web site at www.alsa.org.

Are There Alternative Treatment Options?

N

ontraditional treatments such as 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. Alternative practitioners may provide very sincere support and can function as members of your ALS care team.

When thinking about any alternative treatment for ALS, consider the following issues:

1. What claims are being made?
2. Is there scientific data to support the claims or only testimonials?
3. Are there any peer-reviewed publications?
4. What is the scientific theory or rationale behind the treatment?
5. What is the cost?
6. Would you have to travel a longer distance and are multiple trips required?
7. What are the potential harmful side effects?
8. What advice does your physician offer?

Many ALS patients are using vitamin and supplemental therapies. Several of these have been shown to be of benefit in the transgenic mouse model of ALS. Included are antioxidants such as vitamin E, and supplements such as creatine, ginseng and soy isoflavones. ALS patients should discuss a vitamin and supplement regimen with their physicians.

What Treatments Should You Be Cautious About?

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aving been diagnosed with ALS, a serious and formidable illness, you might be vulnerable to fraud. Be skeptical of claims which promise to revive strength, reverse symptoms, or even cure ALS usually offered (for a high-dollar figure).

There are treatments which take advantage of your sense of feeling vulnerable and may cost your life savings or family assets. The best way to avoid such damaging therapies is to discuss them freely with friends, family, ALS doctors and specialists, and even other patients. Never agree to any treatment under pressure, but take the time to reflect on the pros and cons of each decision prior to committing your time, money, and/or hopes.

The Last Word

Now is a hopeful time for people living and dealing with ALS, because promising new disease-specific treatments continue to be tested and made available. With the aid of these new medications, you can look forward to living longer and maintaining your quality of living along the way.

No two patients will experience ALS in exactly the same way. You must make your own choices and decisions as you live your life. It is up to you. There is no challenge you cannot overcome once you have made up your mind. Many ALS patients have demonstrated how adaptive human beings can be in order to continue living life to the fullest—even in the face of adversity.

This manual has offered a comprehensive look at the various symptoms which may or may not occur in your case, and has explained treatment options to help you improve the quality of your life with ALS.

Notes

Notes

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

Manual 1

What's It All About?

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

Manual 2

Coping with Change

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

Manual 3

Managing Your Symptoms and Treatment

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

Manual 4

Functioning When Your Mobility Is Affected

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

Manual 5

Adjusting to Swallowing and Speaking Difficulties

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

Manual 6

Adapting to Breathing Changes

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

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



27001 Agoura Road, Suite 250
Calabasas Hills, CA 91301
Telephone: 818/880-9007
FAX: 818/880-9006

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

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

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