



The ALS Association

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Member National Health Council and
National Organization for Rare Disorders

The ALS Association
guide for patients
and families

*Basic
Home care
for ALS
patients*





The ALS Association is the only national not-for-profit voluntary health organization whose sole mission is to lead the fight to cure and treat ALS through global, cutting-edge research, and to empower people with Lou Gehrig's Disease and their families to live fuller lives by providing them with compassionate care and support.

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Basic home care for ALS patients

The ALS Association guide for patients and families

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1999

Acknowledgements

We acknowledge with gratitude the work of the original authors Mary Blount, R.N., B.S.N., M.N., and her colleagues Nancy Luttrell, R.N., B.S.N., Phyllis Mayo, R.N., B.S.N., Lynn Muros, R.N., B.S.N., at the University of Virginia Medical Center, Charlottesville, Virginia as well as Lynn M. Klein and Barbara Dolgonos for their important contributions in making this booklet a reality. We also express our appreciation to the many individual ALS volunteers who added their suggestions based upon their person experiences with ALS

PREFACE

One of the major purposes of The ALS Association is to help ALS patients and their families live as full and normal a life as possible.

We believe that the ALS patients and families using this booklet as a guide will be better able to cope with the day-to-day challenges of *living* with ALS.

PURPOSE OF THIS HANDBOOK

The purpose of this booklet is to provide basic information about home care for people affected by ALS. While a variety of symptoms are manifested in individuals afflicted with the disease, a rather systematized progression of ALS is reflected in this booklet.

This book is intended as an introductory guide and should be used along with professional medical care from one's physician, nurse and social worker. More detailed information is available from The ALS Association in the *Living with ALS* manuals. (Further information at the conclusion of this booklet.)

BRIEF OVERVIEW OF ALS

Amyotrophic lateral sclerosis (ALS) is a disorder of motor nerves. For reasons that are only beginning to be understood, these motor nerves degenerate and die.

When the motor nerves can no longer send messages to the voluntary muscles, there is muscle weakness. This might be noted in the arms, legs, speech, swallowing, or breathing muscles. When muscles no longer receive the messages they require to function, they begin to atrophy (waste away). ALS is often referred to as "Lou Gehrig's Disease." Although scientific research is making headway to identify clues to the cause of ALS, there is no cure today.

Three known classifications of ALS have been described:

- *Sporadic* – in about 90% of people with ALS.
- *Familial* – about 10% of people with ALS have a clear family history of the disease.
- *Guamanian* – an extremely high incidence of ALS was observed in Guam and the Trust Territories of the Pacific during and following World War II.

At the onset of ALS the symptoms may be so slight that they are frequently overlooked. The progressive course of the disease might include the following:

- twitching and cramping of muscles, especially those in the hands and legs
- weakness of the arms or legs
- "thick speech" and difficulty in projecting the voice
- swallowing may be difficult
- exaggeration of laughing or crying (greater than your feelings)
- breathing is gradually affected

ALS is unique in each person – in the area(s) of the body affected as well as in the rate of progression. Not everyone develops all of the symptoms.

It is important to stress that ALS generally does not affect thinking and memory. Nor does it interfere with the ability to taste, see, smell, hear or recognize touch. It does not affect function of any organs including the heart.

Much can be offered to manage the symptoms of ALS so that the patient's function is maintained and supported as long as possible. There are medications that help with many of the other potential problems in ALS such as cramps, excessive saliva, and sleep disturbances. Rilutek[®], is the only FDA approved drug at this time. It has been shown in a clinical study to have a positive effect on survival.

DIET AND NUTRITION

Nutrition is an important consideration in the health care of any person and the patient with ALS is no exception. Care should be taken to have a well balanced diet. As ALS progresses, difficulties in maintaining good nutrition may be encountered. The following suggestions may be helpful:

- Eat for enjoyment and to maintain a reasonable weight. Some weight may be lost.
- A daily multivitamin is recommended and adds to good general health. Additional vitamins or nutritional supplements are usually harmless but should be for taken general good health. Vitamins and other “health food” formulas will not make ALS better. Consult your physician when adding any nutritional supplement and evaluate the cost as well.
- Consider supplementing meals with high calorie shakes, or a packaged product such as Ensure, or Carnation Instant Breakfast, if you are not eating enough at mealtimes.
- If eating is tiring, you may find it easier to eat five or six small meals rather than three large ones each day.
- Eating is a very social activity. Keep to your normal routine in terms of when and where you eat and with whom you eat.

Swallowing

Many ALS patients experience some degree of swallowing difficulty at some time during the course of the disease. If you experience coughing or choking with food, in particular with water, some changes are needed. There are several creative cookbooks containing recipes and techniques to expand the variety of foods for easy swallowing. (Refer to *Maintaining Good Nutrition* booklet)

- Give yourself enough time to eat. Don't rush.
- Thin liquids are usually the first to give you difficulty. Water will “go down the wrong way” more often than tomato juice will.
- If a food give you difficulty (such as lettuce), avoid it.
- Softer foods (eggs, mashed potatoes, pasta) require less energy to eat.
- Take smaller bites. Chew and swallow carefully.

- Consider a different method, such as a feeding tube, to get food to your stomach if you have any of the following problems:
 - no appetite
 - too much weight loss
 - eating is no longer enjoyable
 - it takes a long time to eat (more than one hour) a meal
 - eating tires you out
 - food is “going down the wrong way” daily

Self-feeding aids

There are various modifications in eating utensils that may make it easier to feed oneself. Some examples of these modifications include:

- Built-up handles for flatware and utensils are especially useful to people with a weak grasp.
- Plastic cups and glasses are lightweight and easier to hold than glass or china



Check with a rehabilitation or occupational therapist about special devices to assist with self feeding. Product catalogues are available at medical supply stores, from your therapist or nurse.

ELIMINATION

The sphincters that control bowel and bladder functions are voluntary muscles that we learned how to control in early childhood. It is rare in ALS to have weakness of these muscles; therefore, ALS patients generally do not become “incontinent.” But, getting to the bathroom or getting on and off the toilet can be a problem. An elevated toilet seat makes sitting and rising easier. A handrail on the side of the toilet may provide needed stability. A bed side commode is handy for a person who is very weak. Bedpans are not needed for most ALS patients.

Bowel function

The bowel is made of smooth muscle, not voluntary muscle and so the bowel itself is not affected by ALS. However, changes in diet, exercise and fluid intake and weakened abdominal muscles can lead to constipation. You can manage this by:

- drinking enough liquids (6-8 glasses a day)
- get adequate fiber in your diet (bran muffin, breakfast cereal, Metamucil, Fibercon, fruits)
- be as active as possible

If the stool is hard, then you need more fluids. A stool softener such as Colace, may help. You should have a bowel movement at least every 2-3 days. Serious constipation can lead to bowel impaction. Do not let this happen. Talk to your physician or nurse if you are constipated.

Bladder function

Urinary urgency can be a problem in ALS and should be mentioned to your physician or nurse. There is medicine that can help with this problem. A bladder infection can occur if you are not taking in enough liquids each day. If your urine is dark colored, then you need to drink more fluids.

HYGIENE

Bathing and Skin Care

The bathroom is where most household accidents occur, whether you are weak or not. Safety is a main concern. A “home safety evaluation” performed by an expert can be ordered by your physician. This expert will give you advice on how to make your home as safe as possible. Helpful equipment includes:

- handrails
- elevated toilet seat
- toilet and tub handrails
- bath bench with rail and back
- hand held shower
- power bath lift

It is important to bathe regularly and completely. Although “bedsores” are rare in ALS, if you are not able to move by yourself and to change your position every hour or so, then someone needs to inspect your skin daily to be sure there are no reddened areas. Areas most susceptible to pressure sores are the elbows, heels, buttocks, and coccyx.

Reddened areas should be massaged gently with lotion and future pressure on that area limited to one or two hours at the most. Bath oil added to the bath water may prevent drying and itching of the skin. Foam cushions and other devices are available to help reduce pressure.

Fingernails and toenails should be kept short and clean. Toenails should be cut straight across to prevent the development of ingrown nails. Patients who are unable to manage a nail file or clipper can still retain responsibility for nail care if the file is taped to an easily accessible flat surface.

Care of Mouth and Teeth

Oral hygiene is very important for all of us. Teeth should be brushed well at least once daily. An electric toothbrush may be easier to use than a manual one. A lip balm or petroleum jelly can be applied to the lips to prevent drying and cracking. Your physician can order medication and offer advice to help with this problem.

Some patients who have decreased swallowing ability experience problems with drooling the accumulated saliva. Medical supply stores have small sponges (often pink) on a stick for mouth care.

Clothing

Maintaining your regular activities is important for the spirit and emotional health. This includes dressing every day. Some suggestions for easier dressing are:

- Light clothes—weather permitting
- Loose fitting pullover shirts and blouses without buttons. Sweatshirts and jogging attire can be comfortable and fashionable as well.
- Slip-on elastic ties rather than string tie shoes. All footwear should provide support and be slip resistant.
- Slacks or trousers with elastic tops
- Shoe horns with long thick handles

Wearing apparel of all kinds—slacks, shirts, ties, blouses, dresses, shoes—are all available with Velcro fasteners or can be modified.

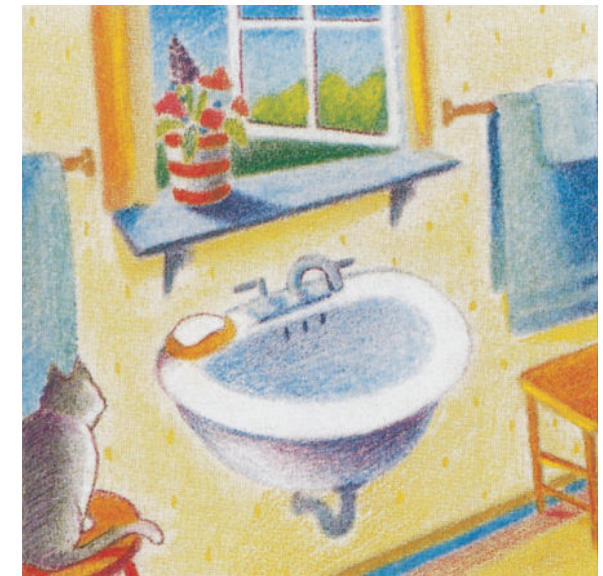
HOME CARE

Home care agencies provide services in the home for people who have medical problems. Your physician must order the specific services. These might include home physical therapy, occupational therapy, nursing care of a home safety evaluation. Most insurance companies pay for “skilled” needs only, i.e., those services that require a professionally trained or licensed person (like a physical therapist). “Basic” or custodial services (such as bathing, dressing, feeding) are rarely a covered benefit. You should check with your insurance company for an explanation of benefits covered in your specific policy.

PHYSICAL THERAPY

Physical therapy can play an important role in your overall well being, both for the body and the spirit. The physical therapist has experience with issues of mobility, exercise, equipment, safety and pain. A physical therapist is able to help in these areas:

- recommend equipment and its safe use
- measure you for a wheelchair and suggest features
- teach you and your family safe techniques for transferring and positioning (wheelchair to car for example)
- perform and teach stretching exercises to prevent or reduce pain
- perform and teach “range or motion” exercises to keep joints mobile and pain free



EQUIPMENT

ALS patients in the latter stages of the disease may require the assistance of special equipment and aids. A variety of aids and equipment that can make you more comfortable are available through medical supply stores. Following is a partial list of such equipment.

- wheelchairs (electric and manual)
- form-fitted seats for wheelchairs
- walkers
- wheelchair lifts and stair lifts
- patient lifts (lifts the patient from one place to another)
- sliding boards
- electric hospital beds
- page turners
- bathroom aids such as raised toilet seats; commode toilet seats
sheepskin (goes under the patient to avoid bedsores)
- folding wheelchair ramps
- external urine catheter for patients confined to bed

The above equipment should be used on the advice of a medical professional. The importance of a hospital bed should be stressed. Changing positions is imperative for the ALS patient. In some instances, when the patient cannot move him/herself, poor circulation, fluid retention, stiffness and bedsores can occur. A hospital bed can alleviate these stresses and prevent discomfort. Hospital beds are available for rental from medical supply houses. A physical therapist, occupational therapist or nurse can give you advice on these and other helpful devices.

RECREATION

Recreation activities become very important to those with limited activity. Mildly affected ALS patients can usually continue to pursue their pre-illness recreational activities. More severely affected patients will need to modify their activities within the limitations imposed by the disease. Some examples of recreational activities suitable for people with limited mobility and significant weakness include:

- Cards: Patients with limited grasping ability may find it easier to use trays that hold the cards for easy viewing.
- Radio/TV
- Reading: Automatic page turners can be purchased or obtained on loan if the patient is unable to do this. If reading is tiring for the patient, talking book services are available in many communities. If these services are not available in your public library, you should request them. Family members may also take turns reading to the patient.
- The Braille Institute Library offers a free talking book service including listening equipment for individuals certified as qualified by a physician. For information call: 1-800-808-2555.
- Playing chess by mail or other board games at home

Of course, when selecting alternative recreational activities, the patient's interests should be kept in mind.

ACTIVITY

Activity is a very important consideration of those with ALS. You should be as active as strength permits but not to the point of too much fatigue. Safety should always be stressed, but it is important to understand that you are the best one to assess your capabilities. The physical therapist can instruct you and your family in any exercises that are considered necessary and beneficial.

Some patients have leg weakness that makes use of a walker advisable. If a walker is prescribed, you should be given careful instruction in its use. A walker with wheels and a seat belt is safest.

A wheelchair should be properly fitted with appropriate back and seat cushions. It is important to have professional guidance before borrowing or buying a wheelchair. Wheelchairs can be purchased with many attachments that can make you more comfortable. Some features are: high back, head support, removable arms, removable leg supports, and tilt. You may need a cane for short distances, a walker for medium distances, and a wheelchair for long distances. It is important, both physically and mentally, to be involved in family and social activities outside your home as well as those in your home.

PAIN

It is often said that pain is not a problem with ALS, but many patients disagree. Actually, then nerve damage in ALS does not cause pain but pain in joints and muscles can occur from reduced mobility. A therapist can teach you and your family exercises that will stretch out those uncomfortable areas. Pain can be well managed and does not have to be a problem. Over-the-counter medications can be helpful. Talk with your physician, therapist or nurse *before* this becomes a problem.

BREATHING

Breathing becomes weak for most people with ALS. The diaphragm is the most important muscle of breathing. The motor nerves that stimulate the diaphragm (and the other breathing muscles) to work will eventually lose their function. This is a gradual and somewhat predictable process.

Breathing problems usually come late in ALS. Early signs may be a soft voice or fatigue with talking. Later, you may become fatigued climbing stairs or with other activities. Some people find that they cannot sleep in a flat position and they start to use several pillows. Tell your physician if you have excessive daytime sleepiness, headaches upon walking in the morning or shortness of breath while lying flat. Your physician may want to test your breathing. A variety of non-invasive (using a small mask or mouthpiece) machines are available that can help you take a bigger breath. These

machines (called non-invasive positive pressure ventilation (NPPV), can give the diaphragm a rest at night so you have more energy during the day.

You should know that there are also breathing machines that can completely take the place of the diaphragm. You usually need to have a tracheotomy (a small hole in the windpipe at the front of your neck) and this type of machine is traditionally called life-support or artificial respiration. The new term is invasive positive pressure ventilation (IPPV) or tracheotomy invasive positive pressure ventilation (TIPPV). Your physician should discuss this option with you and your family.

HOSPICE

Hospice agencies provide services in the home for people who meet all three of the following conditions:

- 1) You have a disease that will shorten your life.
- 2) You do not want artificial life support.
- 3) Your physician certifies that there is some likelihood that the disease will bring your life to a close within the next six months.

Hospice staff is especially trained to have sensitivity to the physical and emotional needs of both you and your family. They are wonderful people who provide both skilled and basic care. They will be a great support for your family.

It is a great relief to patients and family to know that the passing from ALS is peaceful, comfortable and at home for the vast majority of patients. Your physician and nurse will work together with hospice to be sure that you and your family are comfortable.

COMMUNICATION

Many patients with ALS experience difficulty with speaking during the course of their illness. This difficulty may be caused by the inability to project the voice or by the inability to form the words – both of which result from muscle weakness. Speaking more slowly and using shorter words can be helpful. Face the person you are talking with. Family members should resist the temptation to speak to others for you even if they know what you are attempting to say. Most patients do better in a relaxed atmosphere and when they are not being rushed.

There are a number of alternate communication devices and systems available. Some possibilities include a homemade alphabet board, magic slates, and computers. A speech therapist who specializes in “augmentative communication” can make a world of difference to you. Your physician can order a speech evaluation for you.

ADVANCE DIRECTIVES

Every adult should think about what kind of medical care we would or would not want in different situations. It is best to plan ahead so that you do not have to make plans during a crisis. It is normal to become worried after you learn of the ALS diagnosis. Learn how to live with your medical problems and to stay mentally and vigorously engaged.

The first step is to get information about assisted breathing options and discuss it. Understanding something new usually requires hearing about it several times. Involve a family member or friend to help you remember the details. Also, talk to other persons with ALS and their families, or to an ALS support group leader. Read as much as possible and review any available video tapes. Repeated discussions should provide a good understanding of the options, the resources available, and the pros and cons.

Once your plans and wishes are in order, be sure your family and physician(s) know and agree to honor them. Put your health care decisions in writing, in the form of an advance directive to let your family and health care providers know your decisions. Review your decisions from time to time because needs or wishes may change. If you do make changes, make sure the advance directive is updated.

PSYCHOLOGICAL SUPPORT

You are not alone. However, it is normal to feel alone and overwhelmed. Be assured that there are people to help you and your family. Feelings such as denial, anger, withdrawal, and depression are common, normal reactions of both the patient and family members affected by ALS. An atmosphere in which everyone feels comfortable in discussing their feelings will be most supportive. Family members should not give false assurances that “everything will be all right,” but instead, should listen and, in fact encourage the patient to voice his/her fears and concerns and respond honestly. In most cases, with this kind of support, the patient and family members can be helped to reach an acceptance of the illness and prognosis. At the same time, it is important to recognize that acceptance does not mean defeat. Hope is an



integral part of living and should go hand in hand with acceptance. Each patient is individual; the symptoms are individual; and reactions are individual. Neurological research is moving at a faster pace now and no one can predict what tomorrow's findings will bring.

It should be stressed again that in most cases ALS patients have no impairment of intelligence, judgment or other mental faculties. They are still a member of the family and should be treated with the same respect as always, especially with regard to being included in family discussions and decisions. Their opinion should be sought, as before. This attitude will acknowledge that, while their bodily functions are severely impaired, their minds are not. They must be given this recognition that they are alive and a mentally healthy and functioning member of the family system.

In the past few years, many books have been written about helping the patient with a chronic, debilitating illness and the patient with terminal illness. One of the best of these is *On Death and Dying* by Dr. Elisabeth Kubler-Ross which discusses dying and how it is faced by patients, families and the medical professionals responsible for their care. Information presented in this book is also applicable to understanding and dealing with the emotional reactions to chronic illness and could possibly answer many of your questions. This book is widely available in paperback and may be useful to patients and their families.

Another very informative book is *Dying Well*, by Ira Byock, MD, presently director of the Robert Wood Johnson Foundation National Program Office Promoting Excellence in End-of-Life care. It provides a blueprint for families, showing them how to deal with doctors, how to talk to friends and relatives, and how to make the end of life as meaningful and precious as the beginning.

PATIENT SERVICES OF THE ALS ASSOCIATION

The ALS Association National Office provides information about the following services for the patient and family.

Information and Referral Services

- *Medical and Home Health Services* — Referrals are made covering all areas of medical needs from hospitals to nursing homes and in-home care.
- *Information on managing ALS* — Questions are answered or, if necessary, persons are given information on whom to contact for additional data.
- *Psychological Support* — Through a volunteer organization network, referrals are given for counseling and support.
- *Resource Equipment List* — Recommendations and referrals are made on all types of equipment for handicapped persons. Contacts are maintained with manufacturers and suppliers to make sure the most up-to-date materials are presented.
- *Certified ALSA Centers* — Outpatient clinical care facilities providing a multidisciplinary team approach to provide a continuum of care as well as appropriate diagnostic capabilities for ALS patients.

“Living with ALS” Manuals

Six comprehensive manuals explaining day-to-day living with ALS. (See descriptions under “Suggested Readings.”) Also, four companion “Living with ALS” videos that compliment the manuals with equipment demonstrations and further discussion of symptom management to improve living with ALS.

FYI – for your information

Information on a variety of ALS topics.

ALSA's Web Site

ALSA's web site, www.alsa.org, contains information on drug development, information and order forms for the Living with ALS manuals and videos and FYI materials (described above) and other resources for patients and families.

Local Chapters and Support Groups

Throughout the United States, the ALS Association has local chapters and support groups that provide referrals and other patient programs on a local level.

Information and Education for health care professionals

In addition to increasing public awareness of ALS, The Association provides an ongoing information and education program to the health care community to assist them in better meeting the needs of the ALS patient and family.

For information on any of the above services, contact The ALS Association National Office Patient Services Department at (818) 880-9007 or (800) 782-4747 or email info@alsa-national.org.

SUGGESTED READINGS

The following books are available through The ALS Association:

- *Living with ALS* manuals by The ALS Association

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.

Coping with Change

This manual addresses the psychological, emotional and social issues that you must deal with when our 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.

Managing Your Symptoms and Treatment

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

Functioning When Your Mobility Is Affected

This manual covers the spectrum 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.

Adjusting to Swallowing and Speaking Difficulties

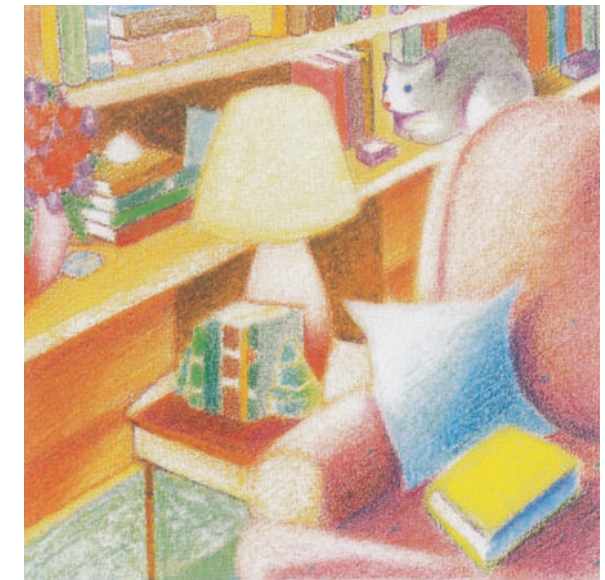
This manual discusses how your speech can be affected by ALS. It covers specific techniques and devices available for improving communication. In addition, swallowing difficulties and how to maintain a balanced diet are covered.

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.

- *Maintaining Good Nutrition with ALS*
from the ALS Association

The ALS Association National Office maintains a reading list of recommended book available through the publisher or book stores. Please contact the Patient Services Department at (818) 880-9007, or (800) 782-4747.



CONCLUSION

It is not possible for this booklet to provide all answers to all questions asked by patients with ALS and by their family members. For this reason, close communication with members of the medical profession is essential. This includes physicians, nurses, physical and rehabilitation therapists, psychologists, psychiatrists, social workers and others.

The ALS Association is a major source of information and support for the ALS patient and family. We care and we want to help you. Patients and caregivers may use our toll-free telephone number: 1-800-782-4747. Please call on us!

Thank you.



A MESSAGE OF HOPE

You can send a message of hope to the hundreds of thousands of people who have ALS or will be diagnosed with this devastating disease.

Today many give to The ALS Association to support its full-time efforts to eradicate ALS from the face of the earth; to help patients cope with its effects.

But, what about tomorrow? Your gift, bequest or memorial can be a message of hope in conquering ALS, to discover its cause and cure. It can be a gift of life to future generations.

If you wish to receive information about making a bequest through your will or have any questions regarding planning or making memorial gifts, we invite you or your attorney to contact The ALS Association Development Department. See back cover for address and telephone numbers.

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1999, 2002.