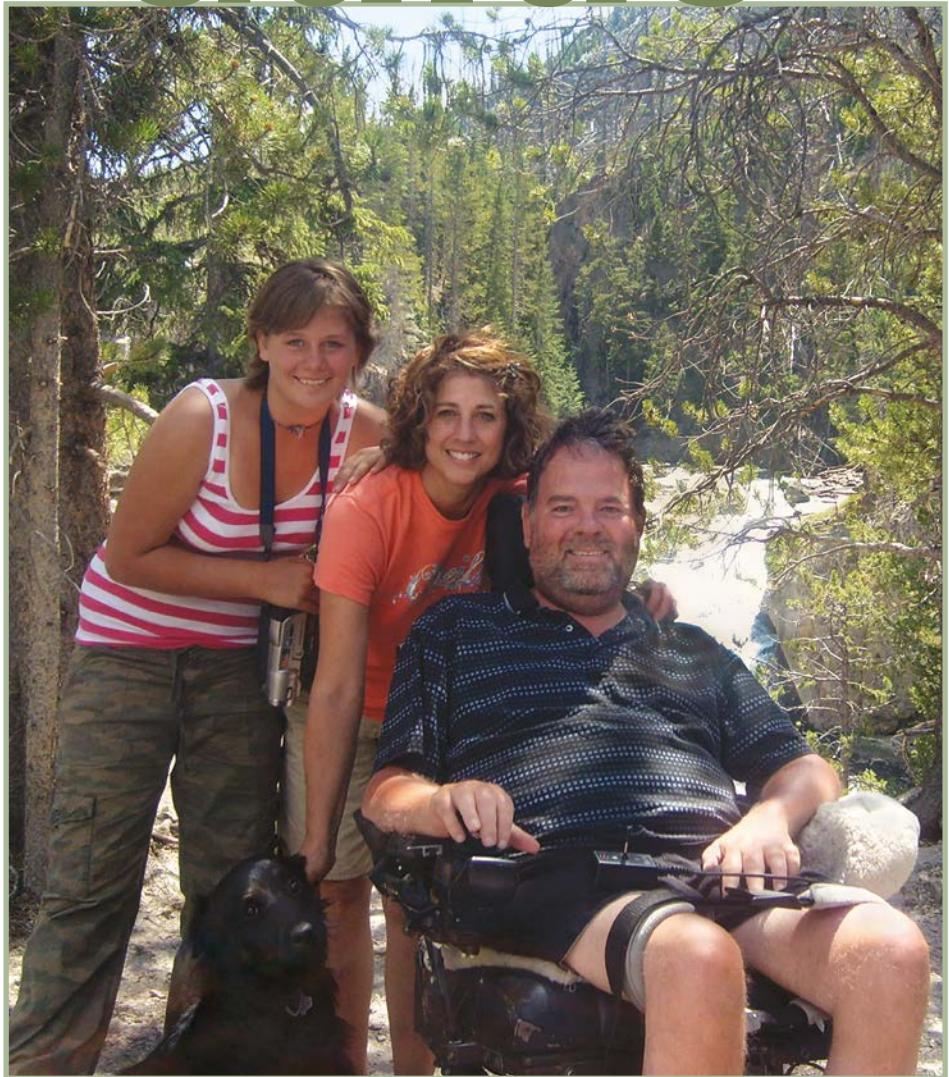


MDA ALS

Caregiver's Guide



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ALS DIVISION

MDA ALS Caregiver's Guide

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MDA welcomes comments and suggestions on this publication.

Acknowledgments

This book is a companion piece to MDA's publication, *Everyday Life with ALS: A Practical Guide*. While *Everyday Life* is written for the person with ALS, and focuses on techniques and equipment for daily living at various stages of the disease, this book is addressed to the family caregiver, whose experience of ALS is certainly as intense and all-consuming as that of the person with the disease. Together, these two books are intended to guide families living with ALS to answers to many of the questions that arise in that experience.

The *MDA ALS Caregiver's Guide* is most indebted to those caregivers who've corresponded with us, participated in Web discussions and been interviewed for articles in the MDA/ALS Newsmagazine. Their candid sharing of their personal experiences, questions and emotions guided MDA's staff in deciding what issues should be addressed in this book.

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The quotations used throughout the book are all from ALS caregivers or people with the disease. Unless otherwise credited, they were taken from previously published MDA materials or from Internet discussions, especially the invaluable Living with ALS forum at health.groups.yahoo.com/group/living-with-als.

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The MDA Publications team

ALS Division — MDA

MDA is the world leader among voluntary agencies in fighting ALS (amyotrophic lateral sclerosis, or Lou Gehrig's disease). Since the early 1950s, when Eleanor Gehrig served as MDA's national campaign chairman, MDA has assisted those affected by the disorder named for her husband, baseball great Lou Gehrig, who died of ALS in 1941 at the age of 37.

MDA's ALS Division offers the most comprehensive range of services of any voluntary health agency in the nation, and leads the search for treatments and cures through its worldwide research program.

For more about MDA's ALS Division, visit als.mda.org.



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Introduction

MDA ALS Caregiver's Guide



The Journey

If you're reading this guide, it's probably because someone you care about deeply has ALS.

This guide will help you to help your loved one through the progression of the disease. It also will help you with your own journey as an ALS caregiver.

Being a caregiver for a person with ALS is emotionally and physically strenuous, but also deeply rewarding in highly personal ways. It's different from caring for a person with any other disease, and may be far more emotionally and physically demanding than other caregiving journeys. ALS caregivers have called the experience life-changing and found that it strengthens character, deepens compassion and brings relationships to new levels of love and trust.

And it's really hard work. It involves learning about new tasks and equipment, while going through the emotions that arise when a loved one has a progressive, debilitating illness. These pages contain practical and emotional strategies for being an effective caregiver. The **MDA ALS Caregiver's Guide** is meant to give guidance, assistance and advice in many aspects of caring for a person you love who has ALS.

It's full of references to other websites, publications, articles or organizations where you can go to find out more about a specific topic or product. And it's laced with quotations from other caregivers and people with ALS, meant to show you that others have dealt with everything you're facing, and they've felt the same fears, despair, exhaustion and hope that you'll be experiencing in this part of your life. You'll read many comments from people about the happiness and satisfaction they've found while living with ALS.

Resourceful caregivers have come up with ingenious solutions to their challenges. Not every suggestion works for every caregiver; sometimes you'll find a better idea than those mentioned here. Because caregiving is an ongoing, organic, creative process that's a little bit different for everyone engaged in it, this book can't include every possible idea. But we hope it provides a platform from which you can think creatively as well as information to get you started and many places in which to look for more solutions.

“ I will forever treasure the time I was able to spend with my Mom during her illness. I learned so much about strength, courage and grace from her. There were tough times (mostly due to my impatience!) but the good times were extraordinary! ”

“ I was worried about everything that people worry about in this situation. How would I take care of my husband? How would I keep my job? How does this disease unfold? How will I know what to do?

How will this affect our son? Will my insurance be enough? Will we become destitute? Will I be strong enough to support him through this? Will I be physically strong enough to even move him? Will we be able to plan for the future?

All the questions that people lose sleep over were the questions that plagued me day and night. ”

“ It's been nine years since Glenn was first diagnosed. I have changed. I discovered that I have faith in myself. I can trust my instincts, and I believe in my basic self-worth. I understand better who I am and what I'm doing with my life, and that is what really matters. ”

— **Caregiver's Reprieve**
by Avrene Brandt, Impact Publishers, 1998

“ As caregivers, we have the potential to burn out both emotionally and physically. My biggest piece of advice is: Don't! I wore myself out, and that caused my husband more anguish. ”

“I'm probably the most fortunate man in the world to have such a wonderful, wonderful wife who's given her life in raising our family and in helping other people so much. Now I have an opportunity to return to her all the kindnesses that she's done for our family in taking care of her. **”**

“I'm not "saving" anything; we use our good china and crystal for every special event such as losing a pound, getting the sink unstopped or the first amaryllis blossom. **”**

“To be able to care for someone or to be needed is a pretty good high. There were good times and a few bad times — usually 3 a.m. and no matter how I placed the pillows he wasn't comfortable. The bad times were few and far between. We caregivers are the lucky ones: You will never be a burden, let us have the good feeling by taking care of you. Besides, maybe we will find the cure soon. Every day is one day closer to the cure. **”**

The primary family caregiver is the care coordinator for the loved one with ALS, the manager of the loved one's medical, social, financial and cultural life. The issues you'll face as primary caregiver will challenge you and your family in ways you've probably never before encountered. You'll make many personal decisions that will affect all of those close to you and your loved one. Along the way, you'll interact with health care professionals, agencies, insurance companies and many other entities.

A good way to start your journey might be to hold a family meeting (including the person with ALS) and discuss who can cover which caregiving tasks, and how care for your loved one with ALS will be coordinated. One person — usually the spouse, but sometimes a parent or an adult child, sibling or other loved one — becomes the primary caregiver. Other caregivers should treat this person as the leader, the one who coordinates and keeps track of the loved one's many needs. Those needs will change, sometimes rapidly, so more tasks will need to be assigned and your list of caregivers and helpers will grow. From the beginning, think of caregiving as a group endeavor, not something to be done by a single person, — otherwise, burnout and collapse are more likely.

No doubt you've heard it said you must "care for the caregiver." This isn't an empty phrase. You must find a balance that allows you to give quality care to your loved one and still maintain your emotional and physical strength. This means that in the course of your journey, you'll need to find other people to help.

This guide offers strategies for maintaining your strength and finding help.

It frequently refers to MDA's *Everyday Life with ALS: A Practical Guide*. Everyone with ALS who's registered with MDA is entitled to a free copy of *Everyday Life* from their local MDA office. These two books are meant to be used together. *Everyday Life* focuses on practical strategies and equipment, and is written for the person with ALS, though of course its information is invaluable for caregivers as well. *Everyday Life* covers: equipment for daily living, saving energy, home modifications, equipment for mobility, speech and respiratory issues, transfers, and exercise.



In the *MDA ALS Caregiver's Guide*, the approach is a little different. This book is geared to what caregivers need to know, including how to know when some aspect of your loved one's condition requires a new intervention, and provides answers to what caregivers can do at various points for their loved ones with ALS.

Here's a brief outline.

Chapter 1: The ALS Caregiver

What is ALS, and what is the ALS caregiver's role?

Chapter 2: Daily Care of Your Loved One with ALS

A glossary or brief encyclopedia of some 60 terms and topics that are likely to come up in the course of ALS. It provides very practical tips on subjects ranging from bathing to Gulf War syndrome, and extensive references on all topics.

Chapter 3: Respiratory Issues

What are the signs that ALS is affecting breathing? What can be done to improve respiratory function in the early and later stages of ALS? The chapter covers handling coughing, emergencies, assisted ventilation and more.

Chapter 4: Communication Issues

How does losing the ability to talk affect self-concept and relationships with family? There are solutions ranging from paper and pencil to role adjustments and dedicated computers.

Chapter 5: Nutrition Issues

Maintaining good nutrition is important in any condition of health. Here's information on nutrition, choking, feeding tubes, etc.

Chapter 6: Emotions

Everyone in the household and the community of someone with ALS is affected by the disease. This chapter advises on coping with your loved one's emotions, your emotions, the family's emotions, children, extended family and friends, and finding support systems.

Chapter 7: Financial, Legal and Medical Issues

Here are tips and references on finding funds, government assistance programs, insurance, low-cost equipment, power of attorney and more.

Something from Within

It's not feats of strength
that makes you a man
or scoring the winning goal
to the blare of the band,
it's not boasting about
the sixty hours you put in
it's something much more
it's something from within.

The soul of a man can't be tested
on the field or the ground,
a real man is noblest
when no one's around,
when it's silent and dark
the meaning of TERMINAL hits home
although shaken
he won't let her fight that monster alone.

He carries her to the restroom
on a signal only they know
Countless times he'll lift her
Through pain he won't show,
Two o'clock, three o'clock
the saliva is cleared from her throat
Not mechanically
but by a husband working the scope.
Her illness will take her
as he fights for her life
The champions her husband
and his life is his wife,
This is a living and dying battle
that few men will face
the something from within
is called COURAGE and GRACE.

He knows in his heart
if things were reversed
She'd do it for him
Because it's something they share
It's something from within!

— Roger Gomez

Chapter 8: Finding Caregiving Help

Help is available from agencies, friends, families and other sources. What should you ask for, and how can the caregiver find some respite? When do you consider a nursing home or hospice?

Chapter 9: End-of-Life Issues

Read this only when you're ready to face the final stages of your loved one's ALS, not before. As ALS progresses, some medical questions will affect survival. The chapter also looks at planning for the last days, funerals and memorials, and handling grief.

“You, our dearest loved ones with ALS, you are the Teacher. You are teaching us lessons of life. To Live. To take each day and be strong. Never ever feel you are a burden. You are giving a meaning, a purpose to all of us, your families, to understand what it is to truly care, to love and to embrace your gentle beings and to understand. I thank you. **”**

“Suddenly your lives are turned upside down and there is more unknown than known. **”**

“I've been ill this week and not being able to give 110 percent really really scares me. If I won the lottery I'd make a place for all of the caregivers to go to, a warm, dark quiet comfortable spa where we could all go just to rest! **”**

MDA's Fight Against ALS

Always remember that your primary resource on the ALS journey is MDA's ALS Division. The staff at your local MDA office, your MDA clinic or MDA/ALS center can give you sound guidance. The families you meet at clinic and support groups are living this experience with you. Lean on them, learn from others, call them when you need to talk or need help; you'll find others leaning on and learning from you, too.

The Muscular Dystrophy Association's involvement with ALS began in the early 1950s when Eleanor Gehrig, widow of beloved Yankees first baseman Lou Gehrig, was searching for a way to fight the disease that had taken her husband's life. Mrs. Gehrig served more than a decade as MDA national campaign chairman. As of July 2012, MDA has dedicated more than \$307 million to ALS research, services and information programs.

MDA maintains a nationwide network of medical clinics, providing specialized medical services for people affected by any of the more than 40 neuromuscular diseases under MDA's umbrella, including ALS. In addition, a number of clinics are designated as MDA/ALS centers. MDA clinics and MDA/ALS centers are staffed by multidisciplinary teams of health professionals skilled in the diagnosis and medical management of ALS, including symptom control, medical interventions and therapies to help maintain the highest possible quality of life. MDA/ALS center teams may include neurologists, physiatrists, therapists (physical, occupational, speech, respiratory), nutritionists, social workers, pulmonologists, gastroenterologists and medical equipment specialists.

NOTE: For information about the nearest MDA medical services, call (800) 572-1717 or go to mda.org.

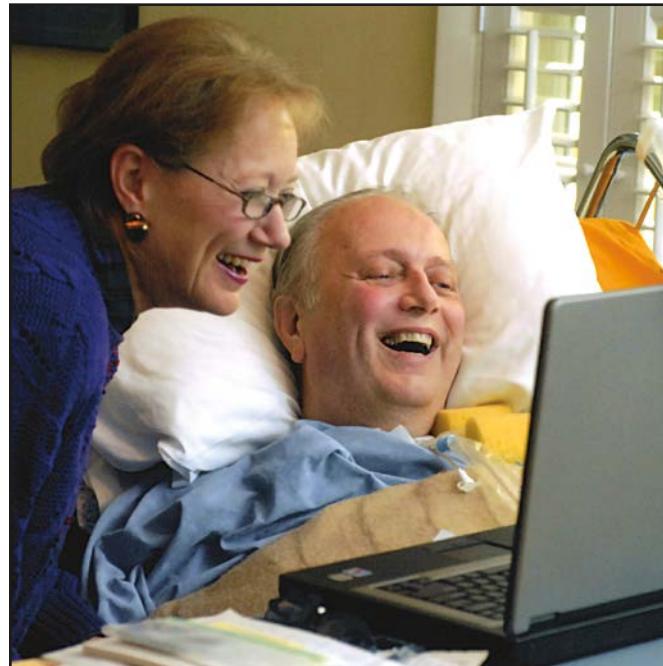
MDA's ALS Division also offers:

- a worldwide program of ALS research that includes basic, translational and clinical research programs;
- the MDA website, mda.org, which contains up-to-date information about MDA's ALS services, clinics and publications; the site also can be used to find the nearest MDA office, clinic or MDA/ALS center, and tells how to participate in local MDA activities;
- an online ALS disease center (mda.org/amyotrophic-lateral-sclerosis) with current, understandable information about ALS causes, symptoms, medical management, research and more;
- assistance with the cost of repairs and/or modifications to all types of durable medical equipment required due to ALS, including wheelchairs, leg braces, communication devices and more;
- a national equipment program that assists individuals with obtaining durable medical equipment, such as hospital beds, lifts, wheelchairs, walkers, shower benches, transfer boards and communication devices; MDA gratefully accepts donations of gently used equipment;
- support and education through support groups, seminars, home visits, community outreach and advocacy, "phone friends" support and Internet chats; and
- publications and videos, available at local MDA offices and on the MDA website (some in Spanish), including:
 - MDA/ALS Newsmagazine online (alsn.mda.org)
 - Quest magazine (quest.mda.org)
 - *Everyday Life with ALS: A Practical Guide* (book)
 - "MDA ALS Division" (brochure)
 - "Milestones in ALS Research" (booklet)
 - "With Hope and Courage: Your Guide to Living with ALS," video for people with new ALS diagnoses and their families

“ Long-term ALS survivors have somebody they love and who loves them. Survivors know they have great value to another person. They feel it's their responsibility to do what it takes to continue to live because someone needs them. ”

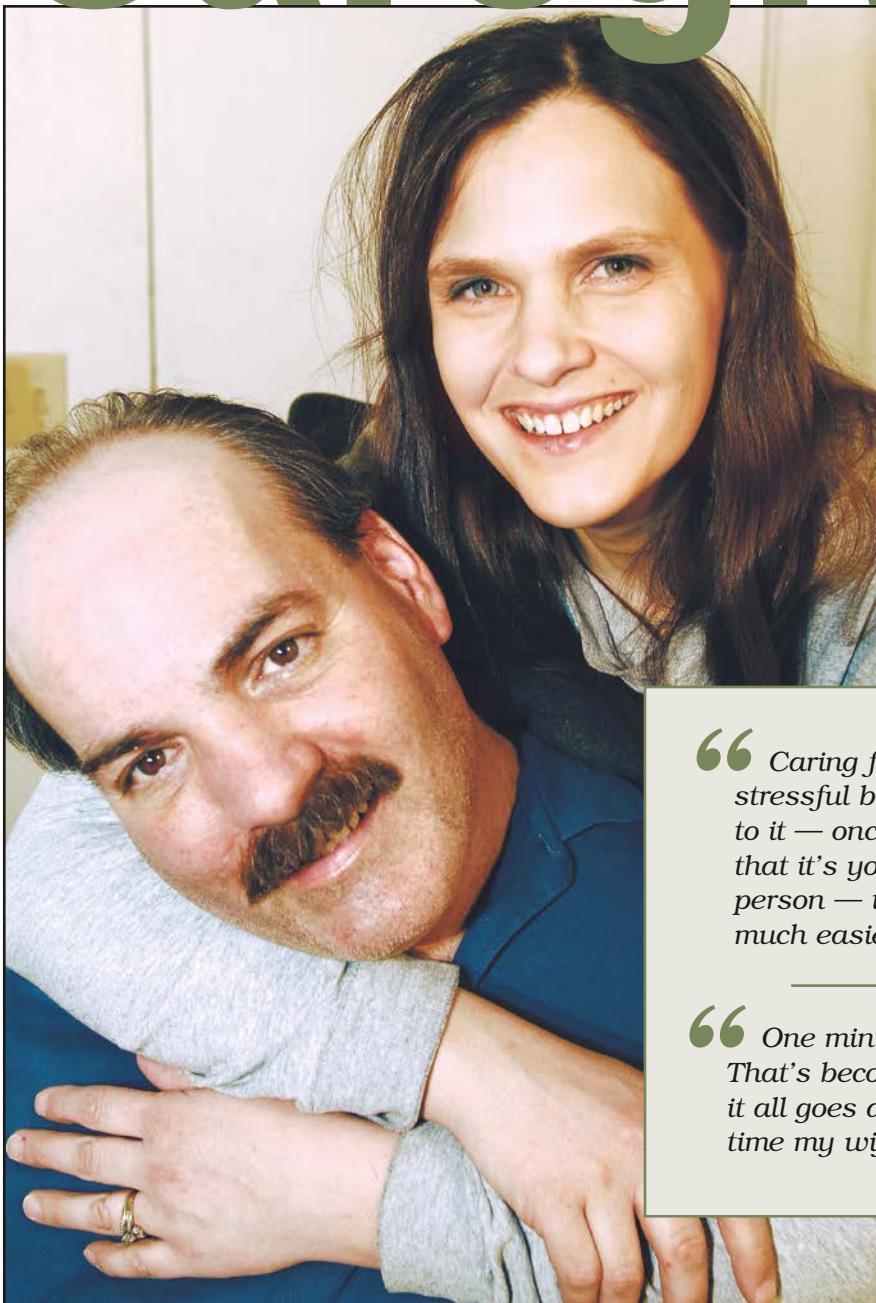
“ Caregivers are our angels on earth. My wife, bless her, has been my sole caregiver for almost 18 years now. ”

“ My wife. I can't even imagine what my life would be like right now without her. She lights up my life in ways she can't even see. ”



Chapter 1

The ALS Caregiver



“ Caring for a person with ALS is stressful but once you put your mind to it — once you make the decision that it’s your life to take care of this person — then the outlook on life is much easier. ”

“ One minute and one day at a time. That’s become my motto. Guilt, fatigue, it all goes away for a little while every time my wife smiles and laughs. ”

What Is ALS?

Amyotrophic lateral sclerosis — ALS or Lou Gehrig's disease — is a disorder of the **motor neurons** or nerve cells in the brain and spinal cord that control the action of voluntary muscles. For unknown reasons, in ALS these motor neurons die, and the muscles they control no longer function, gradually becoming paralyzed.

ALS usually starts out in one of two ways: **bulbar onset** or **limb onset**.

When the motor neurons in the brain stem are affected first, then symptoms start in the face, mouth, throat and tongue, called the **bulbar** region.

More commonly, motor neurons in the spinal cord are affected first and symptoms start in the **limbs** — arms, legs, hands or feet. Rarely, the respiratory muscles are affected first.

In all types of onset, symptoms eventually spread to other areas of the body.

What can you expect?

Each case of ALS is unique.

It used to be that about 70 percent of people died within five years of the onset of symptoms, but that figure is changing thanks to improved medical interventions, like assisted ventilation and feeding tubes. About 10 percent live for 10 years, and some even live for 20 years or more.

ALS doesn't unfold in a predictable pattern. Some people have slow progression of symptoms, while others experience a rapid loss of ability. Sometimes there's a big loss followed by a plateau.

Although each individual follows his or her own course, it's possible to make broad generalizations about the progression of the disease.

Pages 21-23 look at three general stages of ALS: early, middle and late. The time frame and symptoms for each of these stages is totally individual. People with ALS can move very quickly through the stages, or have frequent plateaus and progress slowly, or any combination thereof. The information in the chart gives a general idea of the requirements of life with ALS at each stage.

For the most part, ALS doesn't affect the heart, blood pressure or temperature, or automatic body systems like digestion, urination, defecation, etc. The senses (touch, hearing, vision, smell) are largely unaffected. The muscles that move the eyes, even though classified as voluntary muscles, usually aren't affected, either.

Early Stages

Muscles

- Muscles may be weak and soft or stiff, tight and spastic. Muscle cramping and twitching (**fasciculations**) occurs, as does loss of muscle bulk (**atrophy**).
- Symptoms may be limited to a single body region or mild symptoms may affect more than one region.

Physical effects

- The person may experience fatigue, poor balance, slurred words, a weak grip, tripping when walking or other minor symptoms.
- Sometimes this stage occurs before a diagnosis is made.

Assistance needed

- The person may need help with some physical tasks, and might benefit from the use of a cane, leg brace or other simple device.

Caregiver role

- Provide assistance when asked
- Provide emotional support, including the assurance "we'll get through this together."
- Look for helpful ways to adapt the environment for safety and independence.
- Review and update your legal, financial, medical and other affairs. Investigate government and insurance benefits.
- Begin to share the news of the ALS diagnosis with children, relatives and friends.

Middle Stages

Muscles

- Symptoms become more widespread.
- Some muscles are paralyzed, while others are weakened or unaffected. Fasciculations may continue.

Physical effects

- Unused muscles may cause contractures, in which the joints become rigid, painful and sometimes deformed.
- If a fall occurs, the person may not be able to stand back up alone.
- Driving is relinquished.
- Weakness in swallowing muscles may cause choking and difficulty eating and managing saliva.
- Weakness in breathing muscles can cause respiratory insufficiency, especially when lying down.
- Some people experience bouts of uncontrolled and inappropriate laughing or crying (pseudobulbar affect). Despite how it seems, the person usually doesn't feel particularly sad or happy.

Assistance needed

- **Range-of-motion exercises**, especially for the shoulders, help keep joints limber.
- Leg braces, hand and wrist splints help extremities remain stretched and in a good position.
- Feeding tubes end the fear of choking and keep weight stable.
- Noninvasive ventilation (BiPAP) helps compensate for weak breathing, especially at night.
- Adaptive equipment becomes more valuable.
 - Wheelchairs conserve strength and promote independence.
 - Communication devices enable people to stay part of the conversation.
 - Adaptive eating, writing and computer tools preserve independence.
 - Shower chairs make hygiene easier and safer.
 - Lift chairs and rising toilet seats help with getting up from a seated position.

- Transfer boards and mechanical lifts enable safer, less strenuous transferring from one location to another.

- Anti-depressants and anti-anxiety medication help bolster coping skills.
- Several drugs help with uncontrolled laughing or crying (pseudobulbar affect).

Caregiver role

- Ask for help; don't try to do it all.
- Give your loved one physical assistance with eating, drinking, bathing, toileting, dressing, communicating and mobility.
- Ensure range-of-motion exercises are performed each day.
- Evaluate your loved one's standing, walking and swallowing abilities, and make changes to ensure safety.
- Help obtain needed assistive equipment.
- As much as possible, help your loved one stay involved in the affairs of daily living, including decision-making and planning.
- Watch for signs of depression and discuss them with the doctor.
- Be aware of your own depression and/or anxiety. Discuss it with your doctor and work out coping strategies. Keep on top of your physical health.
- Learn how to operate equipment and perform care tasks from nurses, therapists and others on the health care team.

“To those of you caregivers who have chosen to not only stick by your loved one with ALS but actually love them as partners, not just patients, I commend you. You did have a choice to stay or go ... and you chose to LOVE and STAY. May you know each day that your sacrifices are not in vain. **”**

Late Stages

Muscles

- Most voluntary muscles are paralyzed.
- The ability to move air in and out of the lungs is severely compromised.

Physical effects

- Severely limited mobility. Unable to care for own needs.
- Poor respiration causes fatigue, fuzzy thinking, headaches and susceptibility to pneumonia.
- Speech may be no longer possible.
- Eating and drinking by mouth aren't possible.

Assistance needed

- Power wheelchair, hospital bed, mechanical lift, switches that enable any moving body part to operate computers, environmental control units and communication devices.
- Assisted ventilation, either noninvasive or invasive (tracheostomy).
- Feeding tube.
- Urinary catheters aren't required but can make toileting easier.

Caregiver role

- Find and train caregiving help.
- Oversee 24-hour care of your loved one.
- Find ways to help your loved one stay socially and mentally active.
- Get into a routine that supports you as well as your loved one.
- Although this stage is all-consuming for caregivers, surprisingly many report great stability and satisfaction in their daily lives at this later stage of the disease.

Resources: For a more detailed explanation of ALS, see the ALS disease center at mda.org.

Cognitive (thinking) changes or unexpected behavior are common in people with ALS. Sometimes this is due to damage from the disease (see "Dementia," page 118, and "Pseudobulbar affect," page 56) and other times to factors such as depression or poor respiration.

Sexual function remains normal and there are numerous reports of men with ALS fathering children and women with ALS carrying healthy babies to term.

Role of the ALS caregiver

Taking care of someone with ALS isn't like taking care of a person with an acute illness lasting a few weeks, nor is it like caring for someone with a stable, chronic condition, such as someone who has had a stroke. It's somewhere in between.

The caregiver's job gradually becomes more complex as ALS progresses, meaning you'll have time to learn how to give assistance. The health care team and MDA staff will help you learn as you go.

ALS caregivers move from helping with buttons and balance to providing a steady arm, feeding a meal bite-by-bite, helping with more intimate personal tasks, learning to operate medical equipment and supervising a 24-hour care operation that includes other sources of help.

ALS robs people of their movement, but not their spirits. The toughest — and most rewarding — part of your job may be figuring out how to help your loved one still feel in control of his or her life, even as you provide more and more care.

The Family Caregiver Alliance (caregiver.org) lists these "rules of the road" for caregivers:

- Take it step by step.
- Spend time at the beginning understanding your situation and options, so you don't have to rush into action without a plan.
- A team approach is the key to success. (Your loved one with ALS is a central part of that team.)
- Conditions change along the way and your strategies will shift accordingly.
- Knowledge and confidence come a little at a time.
- You must ask for the support you need.

A Caregiver's Bill of Rights

by Jo Horne

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.

ALS feels like a series of losses. Part of a caregiver's job, for themselves and their loved one, is to find the gains in the losses.

“ My wife and I just decided to take each day to enjoy what we have (which isn't much in worldly terms), each other, our families, the world we live in, and the fact that we have been granted the time we have to do it. ”

“ I gave up a job which I loved, gave up going to lunch with friends, hobbies, going to the mall, etc. But now I know this is my calling. I am the best caregiver my daughter will ever have. ”

- 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 needs 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.

(Reprinted with permission of "Today's Caregiver," caregiver.com)

Caring for the caregiver

You *must* take care of yourself. This may seem impossible when so much attention is required by the person with ALS, but if the caregiver gets ill or is overcome by depression, he or she can't do a proper job of caregiving.

Caregivers make a long-term commitment to a 24-hour-a-day job. Feeling not up to it is a normal, common reaction. But with planning, the job is manageable.

Due to the nature of ALS, it's not uncommon for caregivers to experience sleeplessness, anxiety and depression. Physical symptoms can include back pain, headaches, stomach disorders and weakened immune systems.

Studies show that caregivers develop chronic conditions twice as frequently as others their age. The more hours spent caregiving, the higher the chance of illness.

Nurses and other health care professionals at the MDA clinic can help you find resources to assist with caregiving tasks (also see Chapter 8 and Appendix A).

This book, your MDA ALS support group and your MDA health care service coordinator also can suggest ways to effectively handle the job.

Use these resources — don't be reluctant to admit your frustrations, exhaustion or fears. No one can do it all or do it alone.

Watch yourself to make sure you're getting proper nutrition, as much rest as possible, some exercise and some time for yourself. Acknowledge and accommodate your limitations — back problems, illness, time required by children or elderly parents.

One way to make the caregiving job manageable is by building a caregiving team.

As primary caregiver, think of yourself as a captain with troops to whom you can delegate some tasks. Enlist family members, friends, or members of your church or synagogue.

“To all the caregivers out there: The huge sacrifices you make, just to improve the quality of our lives in any way you can — I know sometimes it is trying, but we do appreciate how much you give of yourself for us.

Your station in life, though it may seem menial at times, is among the highest of human endeavors. Thank you. **”**

Start thinking about your team early, and make a list of people who say, "Let me know if there's anything I can do to help."

Equipment also is a godsend to caregivers. There are numerous mechanical devices to assist with lifting, mobility, feeding, etc. Insurance and Medicare often cover the rental or purchase of these items, or they may be available through MDA's equipment program.

Rewards of caregiving

Despite its many demands, caregiving can be one of the most gratifying, and spiritually and emotionally powerful experiences of a person's life.

The intense closeness with the loved one who has ALS, the chance to discover that you can meet needs and solve problems, the gratitude felt by the loved one — all these enrich the psyche and the heart.

Just as caregiving can take a physical and emotional toll, new research shows that the act of helping can do the opposite — bolster well-being, increase coping skills, provide satisfaction and even strengthen the caregiver's ability to ward off or recover from illness.

But these benefits can't occur when the caregiver is overwhelmed.

Support, respite and perspective will help make the ALS journey one of depth, growth and meaning, instead of only exhaustion, sacrifice and despair.

Your life as an ALS caregiver will be unlike anything you've experienced before. Know that you aren't alone in this experience, and that help is there when you need it.



Rules for the Care and Treatment of Caregivers

by Jeff Lester

As a person with ALS who has been fighting this disease for over 11 years (seven on a vent), I realized early on that if I were to have a somewhat normal life, I'd have to depend on my incredible wife, Lisa, to achieve it. This meant that I'd have to follow some rules about the way I treated her, my primary caregiver. It's essential that caregivers and care receivers have some ground rules about their behavior toward one another.

The rules I'd give to caregivers are: Keep the relationship the same as it was before ALS (don't make it an adult/child relationship). And don't abuse the power you've been given as the other person becomes dependent on you.

Lisa's and my close relationship only partially prepared us for the changes that occurred as I became dependent on her for my most basic needs. Therefore, this transition may be even more difficult for two people who don't start out with a close relationship, especially if the person with ALS neglects his or her role in the relationship. That's why the following rules are for the people with ALS:

Don't take advantage.

When I have to ask for something, I ask myself, "Am I using my condition and others' empathy to get something extra?" For instance, we shouldn't demand a gourmet meal when Spaghetti-Os were OK before, or insist on watching what we want on television (I've had to develop an appreciation for Lisa's soap operas since I'm around all the time now). If we violate this rule, I feel it's entirely appropriate for our caregivers to call us on it.

End pity parties.

Those of us with ALS alone are responsible for our happiness. I don't mean that we shouldn't express our true feelings of frustration or pain about our situation, but we shouldn't wallow in it. If we do, it's right for our caregivers to point it out.

Be useful.

No matter the limitations, there always are ways that people with ALS can be useful, contributing members of our households and world. It's up to us to find out what those ways are, even if it's just listening to those around us and being supportive. Contributing is vitally important because it allows us to see that our lives still have meaning.

Take care of yourself.

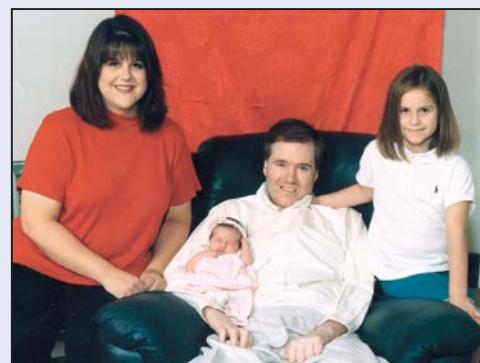
As much as possible, people with ALS need to stay involved in planning for their needs, whether it's a lift, wheelchair, adapted van, special diet, constipation aids, feeding tube — whatever. Also we must keep in mind easing our caregivers' burdens whenever possible. We should keep knowledgeable about the best way our needs can be met, and when possible we should be the leader in our care decisions. It's entirely unacceptable to take a passive, or even worse, a resistant role in care decisions and then complain when something is not done to our liking.

It's inevitable that problems are going to arise between caregivers and care receivers. It's helpful for both to write down their needs and talk together when they're calm. I think the most important thing that my wife and I do is to make sure my care doesn't interfere with our normal spousal relationship. We absolutely don't let disagreements over my care have any impact on our marriage. Care and marriage are best dealt with as two totally separate relationships.

I feel strongly that those of us with ALS have responsibility for our own care and actions, even though our lives may depend entirely on the efforts of our caregivers.

May God bless our caregivers for all they do for us!

(Excerpted
from the MDA/
ALS News-
magazine,
February-
March 2005)



Resources

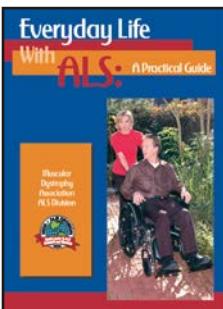
MDA

MDA resources are available from local MDA offices or can be found online at als.mda.org. Also be sure to visit mda.org and click on the “Help Through Services” tab. The Services section offers caregiver information pages, including an extensive list of resources.

Everyday Life with ALS

MDA book about equipment, therapies and accessibility.

“MDA ALS Division” brochure
Overview of MDA ALS services and research programs.

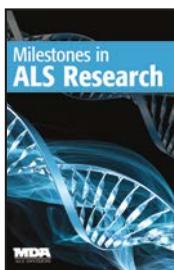


MDA ALS Division website
als.mda.org.

MDA/ALS Newsmagazine online (alsn.mda.org) covers ALS research, medical, psychological, financial and caregiving issues.



“Milestones in ALS Research”
Booklet tracing major research developments since the 19th century. MDA has led the worldwide ALS research effort since the 1950s.



Quest (quest.mda.org).

MDA’s quarterly national magazine covering a range of issues related to neuromuscular diseases. Free to anyone with ALS who’s registered to receive MDA services; available by subscription to others.



“With Hope and Courage: Your Guide to Living with ALS.”

2008 video to orient and inform people with new ALS diagnoses and their families.



Books

Amyotrophic Lateral Sclerosis: A Guide for Patients and Families, 3rd edition, by Hiroshi Mitsumoto, M.D., and Theodore Munsat, M.D., Demos Medical Publishing, 2009

Amyotrophic Lateral Sclerosis, ed. by Robert H. Brown, Jr., M.D., Vincent Meiners, M.D., and Michael Swash, M.D., Blackwell Science, 2000

Amyotrophic Lateral Sclerosis, by Robert G. Miller, M.D.; Deborah Gelinas, M.D.; and Patricia O’Connor, R.N, American Academy of Neurology Press and Demos Medical Publishing, 2005

Biographies of people with ALS

Challenging Nature Photography, by Angelo Sciulli (a person with ALS), 2003

Closing Comments: ALS: A Spiritual Journey into the Heart of a Fatal Affliction, by Brian Smith, Clements Publishing, 2000

Confronted With Death, by B. J. Bloch, Novel Books, 2001

Down the Yellow Brick Road, by Joseph J. Conte and Constance J. Schneider Conte, Tate Publishing & Enterprises, 2006

Facing the Lion: How to Cope with Misunderstanding, Injustice and Suffering, by Pamela Vack, WinePress Publishing, 2001

Falcon’s Cry: A Desert Storm Memoir, by Major Michael Donnelly, USAF retired (a person with ALS), with Dennis Donnelly, Praeger, 1998

Falling Practice: What Illness Teaches Us, by Karen Jorgensen, Silent Press, 2005

I Choose to Live: A Journey Through Life With ALS, by William Sinton, Banbury Publishing, 2002

I Remember Running, by Darcy Wakefield, De Capo Press, 2005

Learning to Fall: The Blessings of an Imperfect Life, by Philip Simmons, Bantam Books, 2003

Luckiest Man: The Life and Death of Lou Gehrig, by Jonathan Eig, Simon & Schuster, 2005

Morrie: In His Own Words, by Morrie Schwartz, Walker Publishing, 2008

On Any Given Day, by Joe Martin and Ross Yockey, John F. Blair, 2000

Tales from the Bed: On Living, Dying and Having It All, by Jennifer Estess, Washington Square Press, 2005

Terminally Sane: One Man's Journey Beyond Lou Gehrig's Disease, by Jay A. Rouelle, Jarou Publishing, 2001

Tonight at Noon: A Love Story, by Sue Graham, De Capo Press, 2003 (about jazz bassist Charles Mingus)

Waking Up: Climbing Through the Darkness, by Terry Wise, Pathfinder, 2004

You're Not You (fiction), by Michelle Wildgen, St. Martin's Press, 2006

Websites, chat groups and listservs focused on ALS

MDA ALS Division
als.mda.org
(800) 572-1717

"Living with ALS" chats are held frequently; check the chat calendar at mda.org/chat/calendar.html.

ALS Advocacy Community
Turning Points in ALS
tpals.org
Stories from people with ALS and caregivers about turning points in the disease.

ALS Care
(336) 340-6400
alscare.com
ALS Care offers telephone consultations with a registered nurse, specializing in ALS home care and other related issues.

ALS Forums
alsforums.com
Publications and extensive chat schedule.

ALS Therapy Development Institute forums
als.net/forum

Caregivers for ALS
z4.invisionfree.com/CaregiversForALS/index.php

Living with ALS Yahoo group
health.groups.yahoo.com/group/living-with-als
Very active forum for people with ALS and caregivers, offering detailed information and support.

NeuroTalk Communities — ALS
neurotalk.psychcentral.com

Patients Like Me
patientslikeme.com/als/community/als

World Federation of Neurology/ALSUntangled
wfnals.org

The role of the ALS caregiver

Books on caregiving

Caring for Yourself While Caring for Your Aging Parents: How to Help, How to Survive, by Claire Berman, Holt Paperbacks, 2005

Counting on Kindness: The Dilemmas of Dependence, by Wendy Lustbader, Free Press, 1993

Candlelights for the Family Caregiver's World: Spiritual Keys for Avoiding Caregiver Burnout and Promoting Inner Peace, by Alice Johnson, Ed.D., and John Johnson Jr., Ed.D., Mal-Jonal Productions, 2001

Caregiver's Reprieve: A Guide to Emotional Survival When You're Caring for Someone You Love, by Avrene L. Brandt, Ph.D., Impact Publishers, 1997 (out of print)

Caregiver Daily Journal, by Sylvia Barron Baca, lulu.com, 2010

Caregivers Are People Too: A Primer for Those Who Take Care of Disabled Persons, by Gloria M. Sprung, M.S.W., Author House, 2005

A Family Caregiver Speaks Up: "It Doesn't Have to be This Hard," by Suzanne Geffen Mintz, President of the National Family Caregivers Association, Capital Books, 2008

The Fearless Caregiver, ed. by Gary Barg, Caregiver Media Group, 2001

If Only I'd Had This Caregiving Book,
by Maya Hennessey, Author House, 2006

The Resourceful Caregiver: Helping Family Caregivers Help Themselves, by the National Family Caregivers Association, Mosby Lifeline, 1996

To Survive Caregiving: A Daughter's Experience, A Doctor's Advice on Finding Hope, Help and Health,
by Cheryl E. Woodson, M.D., Infinity Publishing, 2007

Caregiver-specific websites

AARP

(800) 687-2277

aarp.org

Search "caregiving" for several publications and other resources on aspects of caregiving.

Administration on Aging

National Family Caregiving Support Program

aoa.gov/AoARoot/AOA_Programs/HCLTC/
Caregiver/index.aspx

Caregiver.Com

(800) 829-2734

caregiver.com

caregiver911.com

Produces Today's Caregiver magazine, the Sharing Wisdom Caregivers Conferences, topic-specific newsletters and online discussion forums.

Family Caregiver Alliance

(800) 445-8106

caregiver.org

A public voice for caregivers with programs of information and support at national, state and local levels.

Family Caregiving 101

familycaregiving101.org

The site includes a message board for family caregivers.

Friends' Health Connection

(800) 483-7436

48friend.org

Friends' Health Connection links people with illness or disability and their family caregivers with others experiencing the same challenges.

Mather Lifeways

(800) 492-7500

matherlifeways.com/re_empoweringfamily.asp

Offers a variety of supports for caregivers, especially related to aging.

National Alliance for Caregiving

Family Care Resource Connection

caregiving.org

National Caregivers Library

(804) 327-1112

caregiverslibrary.org

The library, which offers articles, forms, checklists and links, has an ALS section.

National Family Caregivers Association

(800) 896-3650

nfcacares.org

The leading U.S. organization supporting the more than 50 million Americans who care for loved ones with a chronic illness or disability or old age. Offers publications, information, referral services, caregiver support and advocacy.

Rosalynn Carter Institute for Caregiving

(229) 928-1234

rosalynncarter.org

RCI provides educational programs for caregivers, conducts research and disseminates information about caregiving.

Strength for Caring

(866) 466-3458

strengthforcaring.com

A project of Johnson & Johnson, the site offers multicultural resources, including a caregiver manual, message boards and tips for daily living.

Well Spouse Association

(800) 838-0879

wellspouse.org

Chapter 2

Daily Care of Your Loved One with ALS



“ I was totally shocked at how much teaching I would have to do. The student nurses, the full-fledged nurses, all the CNAs, aides, hospital PTs, you name it. I had to enlighten them on ALS and teach them how to use the gait belt properly. I had to explain what happens to your body with ALS. Most were very receptive and wanted to learn. ”

Knowing What to Do

No guide can anticipate all the issues that may come up in day-to-day ALS care, but this chapter offers advice and resources for handling some of the more, and less, common problems that may arise.

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

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Accessibility at home

Home modifications make it easier on everyone. Plan ahead and make modifications before they're needed. Early modifications include taking up anything that can cause tripping, such as scatter rugs or pet toys, and installing handrails in the shower/tub. Later modifications include ramps to the outdoors and increased bathroom space. In two-story houses, a downstairs bedroom space may be necessary.

Modifications range from simple to elaborate: moving furniture, changing to wider-opening door hinges, installing an electric door opener, remodeling a bathroom, installing overhead tracks for an electric lift system (see "Lifts," page 48), building a new bedroom/bathroom suite, etc. Modifications may be tax deductible to some degree.

For more information:

Ask your MDA clinic about getting a home safety evaluation by an occupational or physical therapist.

Everyday Life with ALS, MDA — Chapter 1: Equipment for Daily Living; Chapter 2: Saving Energy; Chapter 3: Home Modifications

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

"Jeanine Schierbecker — Physical Therapist," MDA/ALS Newsmagazine, December 2005

"ECUs Can Help You Take Control at Home," MDA/ALS Newsmagazine, May 2005

"Home Rehabilitation Services: OT/PT Evaluation," MDA/ALS Newsmagazine, October 2002

The Accessible Home: Updating Your Home for Changing Physical Needs, Rockport Publishers, 2003

Adapt My World: Homemade Adaptations for People with Disabilities, by J. Rose Plaxen, Seven Locks Press, 2005

Complete Guide to Barrier-Free Housing: Convenient Living for the Elderly and Physically Handicapped, by Gary D. Branson, Betterway Books, 1991

Universal Designed "Smart" Homes for the 21st Century, by Charles Schwab, AIA, Charles Schwab Architects, 2004

“When I made my first trip out in my power wheelchair with my sister, she got the last accessible parking space at the mall.

As we were laughing about her struggle to release my chair from the van-locking mechanism, an elderly gentleman approached and proceeded to scold and curse us for taking the last handicapped spot, saying he deserved it more since he was much older than I. To which my sister yelled, ‘Oh yeah? Well, we’re more handicapped than you are!’

So much for a smooth first outing. ”

Accessibility outside the home

While the Americans with Disabilities Act (ADA) requires that places open to the public be accessible to people with disabilities, this is not always the case. Whenever possible, check accessibility ahead of time and be ready to be flexible. Notify inaccessible businesses about your problems with access. Get a permit for using handicapped parking spots.

For more information:

["Can't Get In? Work It Out — Accessibility," MDA/ALS News-magazine, April 2005](#)

Americans With Disabilities Act Hotline
(800) 514-0301
ada.gov

Disability Business Technical Assistance Center (DBTAC)
(800) 949-4232
adata.org

Institute for Human Centered Design
(617) 695-1225
adaptenv.org

Alternative therapies

Nontraditional treatments for ALS — such as vitamins, supplements, acupuncture, electrical stimulation and heavy metal chelation — currently lack solid scientific proof that they work. Different people react differently, so an alternative therapy may be worth a try, but proceed with caution.

The important thing is to integrate alternative therapies into conventional medical care. Consult your primary care or MDA ALS doctor before starting anything new. If a treatment won't cause harm, many physicians are willing to work with you in giving it a try. Alternative therapies usually aren't covered by insurance and can be pricey.

Anything that's touted as a miracle cure or that requires a lot of money upfront should be treated very skeptically — or ignored.

For more information:

Memorial Sloan-Kettering Integrative Medicine Service
mskcc.org/aboutherbs

National Institutes of Health National Center for Complementary and Alternative Medicine
nccam.nih.gov

QuackWatch
quackwatch.org

University of Texas M.D. Anderson Cancer Center Complementary/Integrative Medicine/Education Resources
mdanderson.org/departments/cimer

Bathing

As ALS progresses, bathing techniques change. Early on, handrails, shower chairs and handheld showerheads make it possible for people to continue bathing themselves. In middle stages, more help is needed, especially with limb-onset cases. Later the caregiver is completely overseeing personal hygiene, including tooth brushing, hair combing, shaving and nail trimming.

“ My husband would get a daily sponge bath with nice warm water and body soaps. One of us would hold him on his side for the nurse or me to wash his backside.

He loved his baths. It also is a form of mild range-of-motion since we were moving the arms and legs. We used big towels rolled lengthwise up next to him while he was on his side to catch any excess moisture during his bath. We patted him dry, then we would rub him down with a good body lotion.

His skin was beautiful and everyone including his doctors would comment on how wonderful his skin was. **”**

Mechanical lifts and a shower chair with back and arms make the task easier. Usually a person with late-stage ALS needs a sponge bath, not a full bath, daily or every other day. A bathing sling is used on a standard lift and has a cutout for easier transfer into the tub.

A few other tips: Too-hot water and too-vigorous toweling off can cause dry, itchy skin. Similarly, a hot tub or spa may be inadvisable because the heat can overtax the breathing system, and jets can damage weak muscles. For sponge baths, a height-adjustable bed, such as a hospital bed, eases the strain on caregivers' backs. Look for a no-rinse shampoo or a shampoo cap that can simplify the task.

Assistance with bathing and daily hygiene can be found through home care aides, Medicare home health care and hospice staff.

For more information:

See "Lifts," page 48; "Skin," page 56; and Chapter 8. Talk with staff at the MDA office or clinic. A visiting nurse can be hired to demonstrate efficient bathing techniques.

Everyday Life with ALS, MDA — Chapter 2: Saving Energy; Chapter 3: Home Modifications

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

"Splish Splash: Easier Ways to Get Clean," *Quest*, January–February 2008

Blood clots

People with ALS may be at risk for **deep vein thrombosis** (DVT), a type of blood clot that forms when people are immobile. DVTs are extremely dangerous because they can break off and travel to the lungs, causing a life-threatening **pulmonary embolism**. Range-of-motion exercises can help to prevent DVT. Your doctor may recommend taking an aspirin daily or wearing elastic stockings.

Signs of a clot in the legs include: redness; heat or discomfort in one leg; one leg that's more swollen than the other; swelling that doesn't go down overnight; or pain upon standing or stretching calf muscles. Symptoms of a clot that's traveled to the lungs include sudden-onset chest pain and shortness of breath.

If you suspect a blood clot, **don't massage the area**. Contact a doctor immediately. See "Swollen extremities," page 59.

For more information:

"Don't Let Leg Swelling Go Untreated," *MDA/ALS Newsmagazine*, February 2004

Cognitive changes

Approximately half of all people with ALS won't exhibit any cognitive or behavioral disruptions throughout the course of their disease.

The other half will exhibit *some* cognitive/behavioral changes at *some* stage in their disease. In most cases (but not all), the signs will be extremely subtle.

Frontotemporal dementia (FTD) occurs because of changes in the frontal and temporal lobes of the brain, which govern the higher thought processes that make up "executive function." Such processes include: making or following complicated plans, solving complex problems, following a series of directions and making sound judgments. People with diminished executive function may have varying levels of difficulty completing tasks that require complex planning, forethought or organization.

FTD-associated changes in behavior can include: acting inappropriately in public or toward loved ones and caregivers; loss of motivation (apathy); diminished recognition and response to the feelings and needs of others; repetitive or ritualistic activities or habits; and a change in diet

“ [My husband] suffered from frontotemporal dementia. Some days he was right as rain, other days he was totally confused, calling me Mom, not knowing our kids or friends, hearing things, highly agitated.

I used to take a time-out and walk out on the patio and take a few deep breaths. Then I would go back to him, put on the radio and offer him a massage, cream on his hands, a cool cloth and a hug, whatever, to distract him. Most of the time it worked. **”**

that can include new “favorite” foods or eating too much at one sitting.

Other changes, such as lack of interest, introversion or irritability, may be signs of depression ; a doctor can help sort out whether depression treatment will help. Respiratory problems also can contribute to disordered thinking. Counseling can help family members cope with changes in thinking and behavior.

Some tips:

- Organizing and simplifying are important. Have a specific place for commonly used things, such as the television remote and door keys.
- Minimize distractions. If someone is trying to communicate via email or on the phone, make sure that the radio or television is off.
- Encourage doing one thing at a time — not talking on the phone while paying bills, for instance.
- Help the person write notes or use a tape recorder to keep track of things.
- Minimize physical discomfort or pain.
- If the person is stuck on something that’s upsetting him or her, that’s one time when a distraction is a good thing, to divert attention from the upsetting subject.
- Don’t try to convince the person that he or she has changed by pointing out all the changes. That’s usually not effective, wastes energy and can lead to a lot of frustration. It’s not that the person with ALS doesn’t *want* to see; it’s usually that they aren’t *able* to see these changes that they’ve gone through.

For more information:

See “Pseudobulbar affect,” page 54; “Respiratory Issues,” Chapter 3; “Dementia,” Chapter 6; and Appendix B, “When the Thinking Parts of the Brain Go Awry in ALS.”

Constipation

This is a very common and frustrating problem in ALS. It may be the result of general immobility; a side effect of some medications; the need for more fiber and liquid in the diet; and/or muscle weakness that makes it hard to bear down to expel the stool.

Constipation isn’t defined by how often a person has a bowel movement, but by whether the stools are hard, dry and difficult to pass. It’s not necessary to have a daily bowel movement, so long as the task can be accomplished without straining.

Common remedies: Gentle dietary fiber is found in raw fruits and vegetables, bran, seeds or high-fiber cereal bars. Bulk or fiber laxatives such as Metamucil or Citrucel are a concentrated form of dietary fiber. Stool softeners, like Colace, keep stools moist and lubricated. Stimulants like Senokot or Smooth Move, an herbal stimulant laxative tea (made by Traditional Medicinals) increase involuntary muscle contractions, moving the stool along more quickly. A daily capful of MiraLax in 8 ounces of water can pull water into the intestines and soften stool. Others swear by a mini-enema called Enemeez (enemeez.com) or the Magic Bullet suppositories. With a physician’s guidance, keep trying until finding the solution that works best for your situation.

Prescription remedies: Ask your doctor. Mestinon, a drug sometimes used to relieve muscle fatigue in ALS, also has a laxative side effect. It may increase fasciculations, however.

Things to consider:

- Always respond promptly to the urge to defecate. Establish a regular bowel routine, where defecation occurs on a somewhat predictable schedule. Consult your doctor or MDA clinic for suggestions on how to establish a bowel routine.
- Fiber and fluids must be taken together. Without adequate fluid, fiber isn’t effective and in fact can aggravate the problem. This also is true for fiber taken through a feeding tube.
- Although there’s a danger of becoming dependent on laxatives, stimulants, suppositories or enemas, this issue isn’t as acute for people with ALS. Regular and

“We tried to handle as much as possible with humor — some days it was either laugh or cry. We decided to laugh. I even had a cheer I would do when he was constipated — ‘You’re the man, if you can’t do it, no one can. Push it out, push it out, way out.’ It didn’t always work but it made it a little easier. **”**

predictable bowel movements that follow the person's normal pattern (every day or every three days or whatever) make life more comfortable and easier for everyone. One caveat: With regular laxative use, increase fluids and be alert for signs of dehydration, such as irregular heartbeat, disorientation, extreme thirst and headache (see Chapter 5 for more on dehydration).

- Proper positioning helps move things along. A squatting position is more effective than sitting upright. Carefully elevate your loved one's feet on a low stool and have him or her lean forward. Provide stability and support as needed. For additional force, a pillow can be hugged to the abdomen or gentle abdominal massage provided. If using a lift sling, ensure the buttocks are the lowest part of the body.
- Assist weak muscles by gently but firmly massaging downward on the lower abdomen for several minutes. Some have reported success by squeezing together the person's buttocks and massaging downward for several minutes, or by lightly circling the skin around the anus (on the outside) to trigger the colon to move.

When constipation is chronic, fecal impaction may result. This is a large mass of dry, hard stool that can't be expelled. Symptoms include abdominal cramping and discomfort. Watery stool may move around the mass and leak out, soiling clothes. Don't mistake this involuntary release as diarrhea and treat your loved one with an anti-diarrheal product.

Left untreated, impaction can be life-threatening and require emergency surgery. Laxatives won't resolve fecal impaction. If suppositories or enemas don't work, the mass may have to be manually removed by a health care

provider. Suppositories may be given between manual removal attempts to help clear the bowel. If you have any doubts, see your health care provider for a diagnosis.

For more information:

See “Toileting,” page 60.

“Regaining the Simple Pleasure of Regularity,” MDA/ALS Newsmagazine, September 2003

Contractures

To remain healthy, joints must be moved through their range of motion on a regular basis. When joints aren't moved fully, as in ALS, a contracture may develop. This abnormal tightening of muscles and other tissues around a joint immobilizes the joint, causing pain when it's moved. In ALS, this is especially common and problematic in the shoulder joint. Because of the pain, the person moves the joint even less, further aggravating the problem. Contractures can develop very quickly as muscles become paralyzed. Physical therapy and range-of-motion exercises are key to preventing contractures. Massage, splints, braces and proper positioning in a bed or chair may help prevent or ease discomfort. Check with your doctor or physical therapist for more information.

For more information:

See “Pain,” page 50; “Positioning,” page 51; and “Range-of-motion exercises,” page 54.

Everyday Life with ALS, MDA — Chapter 8: Exercise

Coughing

See Chapter 3, “Respiratory Issues.”

Cramps

See “Pain,” page 50.

Crying (uncontrolled)

See “Pseudobulbar affect,” page 54.

Depression

See page 36 and 110.

Dressing

While the person with ALS still can dress without assistance, some techniques can help with weakened hands and arms. Attach a pull, like a key chain, to the ends of zippers. A reacher can help pull up pants or move clothing around. Hooks and loops can be sewn onto skirts or pants. The person should sit for as much of the dressing process as possible; when standing, support should be nearby.

Eventually a caregiver's help will be needed. Larger sizes are easier to manipulate, and looser underwear makes toileting easier. Your loved one can wear pants or skirts with the back seam opened, sitting on a towel in lieu of underwear. When the wearer is seated, the clothing looks normal. This makes toileting easier as no undressing is required.

A person with ALS will feel "more normal" getting dressed, and not wearing a robe or pajamas all day. People with ALS tend to get cold and may need warmer clothes, including boots and mittens (see "Temperature," page 60).

For more information:

Dressing Tips and Clothing Resources for Making Life Easier, by Shelley Peterman Schwarz, Attainment Company Publications, 2001



"Dress for Success"

living-with-als.org/Diane/toilet.html

Search "adaptive clothing" online for websites offering products.

Driving

Caregivers sometimes worry that their loved ones with ALS no longer should be driving. Although it's possible to install hand-operated driving controls to compensate for leg and arm weakness, there's no guarantee that these will remain usable as ALS progresses. Set up a driving evaluation with an occupational therapist or the Association for Driver Rehabilitation Specialists to assess needs and abilities. Caregivers can help ease the transition to nondriver status by rounding up a supply of readily available drivers, investigating taxi cabs and accessible public transportation, and scheduling trips that accomplish several things at once. If you're concerned that your loved one is unsafe on the road but won't give up the car keys, talk to his or her doctor.

For more information:

Association for Driver Rehabilitation Specialists (ADED)
(866) 672-9466
driver-ed.org

Drooling

This has been called one of the most annoying symptoms of ALS. The problem isn't making more saliva, but having less ability to swallow it. This can cause choking, as well as skin irritation, frustration and social isolation due to embarrassment.

In most people with ALS, drooling (*sialorrhea*) can be controlled or at least brought to a tolerable level. In some cases, an occupational therapist can demonstrate saliva management strategies, such as head posture, using facial muscles and achieving more frequent swallowing.

Physicians may prescribe medications to reduce (not eliminate) the flow of saliva. Common drugs include atropine sulfate (Sal-Tropine), tricyclic antidepressants such as Elavil that cause dry mouth as a side effect, and the scopolamine patch usually used for motion sickness. If these are ineffective, the more potent drug Robinul (glyco-

pyrrolate) may be tried. It has some potential downsides, such as constipation, urinary hesitancy, impaired potency, production of mucus plugs, and worsening of existing glaucoma or mental confusion.

Next up in the arsenal is botulinum toxin (Botox) injections in the parotid (salivary) gland. It may take several injections over a number of weeks to achieve saliva reduction, and the effects are temporary. If this isn't successful, doctors may recommend radiation of the salivary glands, which also can take several weeks or months to work.

Reducing saliva flow can cause another problem — too-thick saliva, which can be difficult to swallow or cough out. See "Choking," page 88, for strategies to thin mucus. A dry mouth also can hasten tooth decay.

Remember that drooling and medications that reduce secretions can increase the body's need for water.

Caregivers have tried various creative ways to handle the flow of saliva.

- To catch the saliva, fold a terry cloth towel or washcloth three or four times, and secure it below the lower lip, tucking it behind the jaw or fastening it with an elastic band behind the head.
- Make or buy rolled gauze pads (like those used by dentists) and tuck them between the gum and cheek.
- Those with arm mobility can wear terry cloth sweat bands on the wrists, for frequent mouth wiping.
- Buy face masks (such as those used by transplant patients or people with allergies) and line the chin area with a strip of absorbent cloth. Cut out the portion that goes over the nose if it causes discomfort.
- Cover bed pillows with a waterproof pad and a folded towel to catch nighttime drool.
- Suck up saliva with a portable suction machine, similar to those used by dentists.
- The person can grip a folded cloth or paper towel between the teeth to absorb saliva.

Natural saliva reduction strategies include:

- Before bed, gargle and swish with warm salt water, especially under the tongue (some recommend kosher salt).

- Reduce or eliminate dairy, sugar (including artificial) and grain/gluten products from the diet.
- Use a few drops of anise oil in the mouth (available at health food stores).
- Drink warm tea.

Dry mouth

When using medication to dry up saliva, or when using *noninvasive ventilation* (see Chapter 3), mouth dryness and nose bleeds may result. Remedies include increased fluid intake and attaching a humidifier to the ventilator or in the room. Try an over-the-counter dry mouth spray (available in the oral hygiene section) or moisturizing mouthwash, such as Oasis from Sensodyne.

See "Choking," page 88, for information on dealing with thickened secretions caused by dry mouth.

Ears

People with ALS sometimes report a feeling of the ears being plugged, as if they have water in them or need to "pop." Check with a doctor to rule out infection, sinus problems, wax buildup or other medical causes.

In the absence of these problems, it's possible the cause may be weakness of the muscles that maintain tone in the Eustachian tubes connecting the mouth and ear canals. In addition, weakness in the muscles that elevate the nostrils (thus opening airways) can lead to a chronic feeling of being stuffed up.

Possible remedies include "nose openers" used to stop snoring, such as Breathe Right nasal strips. These strips of tape go across the nose and hold open the nostrils. Antihistamines and decongestants sometimes help, and some say they can get their ears to pop by using their CoughAssist machine (see Chapter 3).

For some with ALS, the ears become painfully folded during side-lying. Some caregivers create an "ear pillow" to prevent this problem. Using a piece of foam of the proper thickness to keep the head in alignment (not tilted up too high), cut out a 4-inch hole in the center for the ear, and cover the foam with a pillow case.

Emergency medical information

Collect important medical information in one spot to facilitate doctor and emergency room visits. Keep this information current, and post a copy on the refrigerator, where ambulance crews have been trained to look in an emergency. Information should include:

- The person's name, birthdate, diagnosis and special information (i.e., can't speak, can't move legs, uses letter board, uses BiPAP, etc.)
 - Emergency contacts and phone numbers (note if someone holds durable or health care power of attorney)
 - Insurance information, including identification numbers, addresses and phone numbers
 - Names of doctors and phone numbers
 - Daily medications: times and dosages
 - Adaptive and supportive equipment
 - Allergies
 - Special diets
 - Advance directives and living will
 - An updated history of surgeries and major medical events

Prominently note any special orders such as "Do Not Resuscitate" or "No Tracheostomy" (see "Advance directives," page 148).

For more information:

National Association of Professional
Organizers
(856) 380-6828
napo.net

Personal Records Organizer

(303) 506-5413
proorganizer.com

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Emergency preparation

Hurricanes, earthquakes, fires, power outages, terrorist attacks — ALS brings special challenges to any kind of emergency. Some planning will help the family be prepared.

Before an emergency occurs, contact your state and local government's office of emergency management or local fire department to find out what emergency relief assistance is offered. They'll tell you whom to contact in event of a disaster, where to go, what to have ready. Some of these offices will have special arrangements for people with disabilities. You may need to register in advance for these services.

Emergency Checklist for People with Disabilities:

1. Prepare a medical information list (see “Emergency medical information,” page 39).
 2. Send copies of important documents to an out-of-town contact person, in case the originals are lost or destroyed in a disaster.
 3. Know which shelters are best prepared to accept people with disabilities and special medical needs.
 4. Have a pet care plan. Shelters don’t always accept pets or service animals. Contact your local animal shelter.
 5. Make an equipment plan.



- Have an extra battery for a motorized wheelchair or scooter. Know how to recharge the battery.
- Have a patch kit, can of seal-in-air product, and inner tubes for a wheelchair with inflatable tires.
- Have a lightweight manual wheelchair for backup.
- Have a converter for your communication device.
- Pack a low-tech communication board and preprinted key phrases in your disaster kit.
- Order an adapter kit for your ventilator so it can be plugged into your car or a marine battery.
- Contact your electricity provider to see if they offer a "priority reconnection service" that will help ensure your power is restored as soon as possible. Even with this service, power could still be out for a long time following a disaster.
- Have a list of the style and serial numbers of medical devices.

6. Create a disaster supply kit and escape plan.

A comprehensive checklist for people with disabilities is available from the American Red Cross at redcross.org/www-files/Documents/pdf/Preparedness/Fast%20Facts/Disaster_Preparedness_for_PwD-English.pdf.

In case you must get out of the house quickly, determine the best escape routes and practice them with the family. Remember that you'll need alternatives to some traditional plans. For example, emergency experts advise heading for the basement in a tornado, but these aren't usually wheelchair-accessible. Will you be able to



carry the person with ALS and their equipment downstairs, or should you go somewhere else?

Assign roles: Who will help the person with ALS? Who will carry supplies? Rehearse, drill, look for other problems. Choose more than one exit in case one is blocked.

It's also a good idea to discuss a finalized evacuation plan with other family members who may not live with you, as well as with neighbors, friends and home care aides in case anyone other than the primary caregiver needs to assist.

Keep emergency phone numbers in your wallet and near telephones where they're available to everyone involved in caregiving.

Caregivers who work outside the home should check with supervisors about any emergency plans in effect at the workplace. For example, some places won't let employees leave for home until an "all clear" has been given by local authorities. Find out whether your home health agency has special provisions during an emergency. Will they continue to provide care and services at another location if your loved one needs to be evacuated?

For more information:

["What Will You Do If the Power Goes Out?" MDA/ALS News-magazine, July-August 2007](#)

["Alternate Power Resources," MDA/ALS News-magazine, July-August 2007](#)

["What Will You Do in a Disaster?" Quest, June-July 2007](#)

["Plan Ahead for Emergencies," Quest, September-October 2005](#)

["Are You Prepared for an Emergency?" MDA/ALS News-magazine, June 2005](#)

[The Home 911 Emergency Info-Book, Vantage Point Books, 2005](#)

[ADA Best Practices Tool Kit for State and Local Governments \(Chapter 7\)](#)

U.S. Department of Justice

ada.gov/pcatoolkit/toolkitmain.htm

American Red Cross and the Centers for Disease Control and Prevention
Evacuation Planning for Persons with Disabilities and Caregivers (2006)
redcross.org/preparedness/cdc_english/evac_dis-1.html

Institution on Community Integration
Impact magazine, Spring/Summer 2007.
Feature issue on disaster preparedness for people
with disabilities
ici.umn.edu/products/impact/201/default.html

Interagency Coordinating Council on Emergency Preparedness and Individuals with Disabilities
disabilitypreparedness.gov

International Ventilator Users Network
ventusers.org

U.S. Department of Homeland Security
Federal Emergency Management Agency (FEMA)
Hotline (800) 621-FEMA (3362)
ready.gov

Energy/fatigue

ALS leads to muscle fatigue which may manifest as general fatigue. Mental exertion also may be fatiguing because of overall effects of the disease.

Whatever activity a person can do is fine; they should rest when fatigue sets in.

Extreme fatigue may indicate breathing problems, which must be addressed with assisted ventilation. See "Respiratory Issues," Chapter 3. In later stages, fatigue may occur even after a passive activity such as being bathed.

Some people with ALS sleep more hours than usual, even 12 or more hours a day. Be sure daytime naps don't keep the person awake at night. Some doctors are using modafinil (Provigil) to help with daytime sleepiness.

For more information:

Everyday Life with ALS, MDA — Chapter 2: Saving Energy

Equipment

"Get it before you need it" is the mantra of ALS. Some people see using assistive equipment as giving in to the disease, but in fact the opposite is true. Adaptive equipment is like a weapon in the battle against the disease.

Canes, braces, walkers, wheelchairs, communication devices, coughing and suction machines, feeding tubes, lift chairs, hospital beds, alternating pressure mattresses, mechanical lifts and assisted ventilation make it possible to thrive as ALS progresses. Assistive equipment enables greater independence and safety for your loved one, while making it easier for you to be an effective caregiver. Not using this equipment can result in injuries to the person with ALS and the caregiver.

Another reason to get equipment early is that it takes time to order and get approval from insurance or Medicare. If a move to assisted living, nursing home or hospice is being considered down the line, be sure to get all essential equipment first, as Medicare may not pay for it afterward.

“Once the medical equipment started to take over the house, it got tougher. We had to face what was coming. We tried to make light of it and realize the end wasn't going to be tomorrow. The stuff we were bringing into the house was there to help him, not shorten his life. **”**

“By getting the feeding device before she needed it, she was able to get used to it, on her own terms in her own time. Later, when she did have to give up eating, her emotional turmoil was not compounded by having to learn how to use the feeding tube.

We approached everything that way, and I still believe that helped us to feel like we were happening to the disease rather than the disease happening to us. **”**

MDA provides financial assistance with the repairs and modifications of medical equipment, including leg braces, wheelchairs and communication devices. It's possible to borrow equipment through the MDA equipment program; check with your local office.

For more information:

See Chapter 7 for information on help paying for equipment.

***Everyday Life with ALS*, MDA — Chapter 4: Mobility & Support Equipment**

"People with ALS Share Personal Experiences with Life-Enhancing Devices," MDA/ALS Newsmagazine, February 2007

"Accessing & Acquiring Assistive Technology — Some Options to Make Funding AT Purchases Easier," MDA/ALS Newsmagazine, November-December 2006

"Technically Speaking, It's a Good Time to Have ALS," MDA/ALS Newsmagazine, March 2006

"Get Time on Your Side When Obtaining Major Equipment," MDA/ALS Newsmagazine, January 2005

"Plan Ahead for Big Purchases — Communication Device," MDA/ALS Newsmagazine, January 2005

"Equipment Survey Can Help with Planning," MDA/ALS Newsmagazine, April 2002

ABLEDATA

abledata.com

RESNA — Alternative Financing Technical Assistance Project
(703) 524-6686

resnaprojects.org/afp/index.html

State Departments of Vocational Rehabilitation
askjan.org/cgi-win/TypeQuery.exe?902



Exercise for people with ALS

It's not known how much exercise — if any — is valuable for people with ALS. Before beginning an exercise program, consult the doctor or physical therapist about frequency, duration and level of intensity. The goal of gentle exercise in ALS is to help maintain mobility, improve endurance and minimize pain from the effects of muscle wasting. When exercise isn't possible, physical therapy and range-of-motion can help serve the same purposes.

Important exercise guidelines include:

- **Don't overdo it:** Soreness after exercise might be an indication that you're damaging muscle, which isn't good.
- **Think aerobic:** This helps improve heart health and circulation. Possibilities include a stationary bicycle, upper body ergometer, elliptical machines or aquatics, or multiple repetitions of gentle resistance strengthening and stretching exercises.
- **With weights, start small:** Increase only when appropriate and by a very small percentage.
- **Just say no to pain:** Forget the motto "no pain, no gain." This doesn't apply to people with ALS. Focus on maintaining functional strength, endurance and independence.
- **Be flexible:** Don't hold to a rigid program. The person should listen to his or her own body and use common sense.

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

For more information:

See "Range-of-motion exercises," page 54.

Everyday Life with ALS, MDA — Chapter 8: Exercise; Chapter 9: Exercise Instructions

"Supervised Resistance Exercise Slowed Functional Loss in Small Study," MDA/ALS Newsmagazine, September 2007

"Exercise Has Many Benefits for People With ALS," MDA/ALS Newsmagazine, October 2002

Exercises for caregivers

Caregiving, especially lifting, is hard on the back, neck and shoulders. Good body mechanics (like lifting with the legs, not the back) and assistance (using a mechanical lift or helper) significantly reduce the risk of injury and always should be observed. But without proper flexibility and strength in legs, hips, shoulders, abdominal and buttock muscles, an injury can be just one wrong move away.

Regular exercise — say an hour at the gym or a daily walk — is a very good idea for caregivers, with both physical and emotional benefits. Talk with a trainer or doctor about a good program for you. Because it can be hard to find the time for lengthy exercise breaks, consider incorporating several quick exercise periods into daily routines.

The following 30-second stretching and strengthening exercises, performed several times throughout the day, can have a positive cumulative effect. They can be fit into odd moments, like while waiting at a traffic light, cooking or standing in line. Pairing an activity with an exercise — say doing partial squats while waiting for the toast to pop, hamstring stretches during a TV commercial and core muscle strengthening while blow-drying hair — can allow caregivers to improve without even realizing it.

NOTE: Check with your doctor before undertaking any new exercise program.

Flexibility: This is critical to preventing injury during the many caregiving tasks that put the body into unaccustomed positions. Stretch to the point of discomfort but not beyond. Pain or distance isn't the point. Hold stretches about 15-30 seconds, repeating several times. Never bounce to get a greater stretch.

- **Lower back stretch:** Tight hamstrings contribute to lower back problems. Stretch hamstrings by sitting in a chair with your feet on a footrest. Keeping knees straight, lean forward until feeling an easy, not painful stretch in the back of the thighs. Or, bend your knees just enough to allow you to touch your toes.
- **Shoulder stretch:** Scratch your back. Point your elbow in the air and scratch the top of your spine. Next, curl each arm around the front of your neck and scratch over your opposite shoulder. Finally, hold your elbow down by your side and twist your hand back up between your shoulder blades.
- **Neck stretch:** Lower your ear to your shoulder on either side.

Strengthening: Muscle strength is important for lifting and for stabilizing the lower back region. To strengthen without using weights, tighten muscles to their maximum, hold for a count of 10, relax for a count of 10 and do several repetitions a day. Be sure you're stable, either by taking a wide stance or holding onto a secure object.

- **Legs and hips:** Do partial squats while holding a stable object like a countertop. Keeping your back straight, slowly lower as if sitting down. Go down as far as comfortable then back up. For greater effect, use only one leg at a time. When rising from a chair, use only one leg. Hold onto an object for stability but don't pull yourself up.
- **Lower back:** Squeeze buttocks and/or abdominal muscles as tight as possible and hold for a count of 10. To strengthen core muscles, pull your navel, waist and buttocks toward the middle of your body, as if trying to pull your body away from your clothes. Hold, release, repeat.

“ Loads of people have had to give up caregiving due to injury. Then they have to find someone else to provide care, or in some cases find a nursing home. The beauty of exercise is, whatever you can do will help. A little bit here and there all adds up.”

- **Shoulders and arms:** (a) Take a weight such as a shoe or soup can and lift it straight up in the front and to the side, from the waist to over the head. Hold arms straight out to the sides at shoulder height, then bring one arm across in front of the chest, keeping the elbow straight. (b) Force shoulder blades together, then relax, repeating several times. Shrug shoulders up toward the ears then relax.
- **Neck:** Place your palm in the middle of your forehead and gently press, resisting the push with your neck muscles. Repeat on each side and the back of your head.

For more information:

[“Exercises to Do on the Run: Simple Ways for Caregivers to Resist Injury,” MDA/ALS Newsmagazine, October 2003](#)

[Treat Your Own Back and Treat Your Own Neck](#), by Robin McKenzie, Spinal Publications, 2006

Eye care

Some people with ALS have burning or dry eyes, or — just the opposite — watery eyes. Either case can be irritating and even painful. This could be caused by lack of blinking or by air blowing from a respirator. Saline eyedrops, or a warm wet washcloth or cold washcloth compress, may help. An antibiotic may be needed if irritation persists. Natural Tears, an over-the-counter product, is recommended. ALS doesn't affect vision.

Falling

This ALS symptom is of great concern to caregivers. Even when a person can walk, trips and falls can occur without warning, and it may be impossible to use the arms to brace for a fall. Injuries from falls range from minor to major and the recovery period can lead to additional loss of physical ability — the last thing anyone with ALS wants.

The best way to avoid falling is to use assistive equipment. However, many people will consent to using a cane or a folding shopping cart, but balk at using more “medical” devices like a walker or wheelchair out of embarrassment or resistance to “giving in to the disease.” Assistive equipment actually allows more independence and fights the disease by preserving precious strength and energy. Caregivers often try to convince their loved ones to use the appropriate equip-

ment, with varying degrees of success.

Some tips:

- Ensure the home environment is safe by eliminating tripping hazards and adding sturdy handholds at strategic locations. Shield sharp corners on tables and counters and add a shower chair in the bathroom.
- Helping someone up off the floor after a fall can be dangerous for both of you. When possible, use a mechanical lift or recruit a neighbor or family member to help. If you can't get the lift to the person, roll the person onto the lift sling or a sheet and pull to a location where the lift can be used.
- Be careful to use proper body mechanics when lifting. If no help is available, call 911. If you ask the rescuers not to use their sirens, they usually won't. See the chapter on transfers in *Everyday Life with ALS* for more on standing up after a fall.
- Ensure your loved one has a cell phone, emergency assistance pager or other signaling device for falls that happen when you're not around.
- If the person with ALS stays alone for periods of time, ensure rescuers (neighbors or emergency personnel) can get into the house by hiding a key outside or leaving the door unlocked.

“I was just starting to waken from a delicious sleep, when I heard a terrible crash. I looked over and saw that [my husband] was face down, his glasses bent, and there was blood all over the hardwood floor. (Always considerate, he had somehow missed the carpeting.)

The cuts on his face were not serious but there was considerable splatter (thanks, “CSI,” for THAT term). He falls from time to time, so at first, it looked like a matter of checking him out, and perhaps getting the Hoyer lift into play.

But he seemed unusually disoriented, he was snoring, and his face seemed slightly purplish. I called 911. **”**

- Protective equipment like knee and elbow pads and a helmet can make falls less injurious. Lighter-weight shoes are easier to negotiate.

For more information:

[“Lifts,” page 48; and “Accessibility at home,” page 32.](#)

[“Take Falls Seriously to Prevent Further Injuries,” MDA/ALS Newsmagazine, October 2002](#)

[“All Fall Down,” Quest, December 2002](#)

Feet

In addition to exercises recommended by a physical therapist, and/or the use of *orthotics* (leg braces), feet need protection in bed, as heels can develop pressure sores and the weight of blankets and *foot drop* (inability to turn the ankle or toes upward) can cause pain. “Float” your loved one’s heels above the bed using small pillows at the ankles. Prevent feet from flopping down or to the side by bracing them with more pillows. Support the weight of sheets and blankets with a blanket lift at the end of the bed. Blanket lifts, which fit between the mattress and box springs and extend upward to support the bedding, can be purchased from medical supply outlets or made at home. Sheepskin pads, knee or elbow pads under the heels, foam boots and heel pads also are helpful.

For foot drop see the medical care team about *leg braces*, some of which the loved one can sleep in.

By the way, caregivers should protect their feet and knees also. A thick rubber kneeling pad can help a lot if you’re frequently up and down. And watch for repeated banging of knees against furniture when turning or lifting the person with ALS. Knee pads or a new technique may be in order.

For more information:

[See “Swollen extremities,” page 59.](#)

“To keep my husband’s feet from falling to the side, a friend constructed a three-sided box lined with 2 inches of foam. He rests his feet inside the box, which is connected to the bed frame and is adjustable. I tent the blankets and sheets over the box. **”**

Hands

As hands, arms and shoulders weaken, caregivers can seek out various aids to extend dexterity. These include: wrap-around lap desks to support the arms; mobile arm supports to allow both horizontal and vertical motion; lightweight wrist splints; eating utensil holders or specially designed eating utensils such as lightweight large-handled cups and plate guards; key holders, doorknob extenders, light switch extension levers; lightweight reachers; card holders (for playing cards); button and zipper hooks; long-handled sponges; Velcro fasteners on pants and shirts; pencil grips; book holders; speaker phones and more. Consult an occupational therapist to help solve specific problems.

Hand-curling can be managed with hand splints, or by putting a rolled washcloth inside the hand, to keep it in a more natural position and prevent fingernails from digging into palms. Holding small, heated rice bags in the hands for 10 to 20 minutes can make them more comfortable.

Some other techniques to help adapt to changes in hand and arm strength include:

- In addition to range-of-motion exercises for the shoulders and elbows, try the “prayer position” exercise. Put the hands together as if praying and point the fingers up and down; this helps stretch fingers and wrists and releases stiffness in forearms.
- A sling supported with a strap across the back and over the opposite shoulder cradles the arm and can help protect the shoulder joint from uncomfortable stretching.
- Arm rests on chairs may need to be padded in order to be high enough to support weakened shoulder muscles. This is especially true if a pressure relief cushion is put on the seat.

“After losing her speech, my Mom's left hand was the first thing to be affected. Her fingers also curled, ending up in sort of a claw position, and it was very painful when the fingers were moved.

When I put her shirts on, I would cup her hand with mine to make sure the shirt didn't get caught on her fingers and pull them. At the suggestion of the hospice nurse, we rolled up a washcloth and she would hold it so her hand didn't become completely clenched. **”**

- A lightweight splint, similar to those available at the drugstore for carpal tunnel syndrome, supports the wrist, helping to bend the fingers easily, which maximizes the grip, helping with many tasks of daily living.
- For feeding, the person with ALS can support the elbow on a few books or pillows or with a suspension arm sling. Another help is The Arm Thing, an arm lifter invented by a man with ALS and his wife/caregiver. Long straws make it unnecessary to pick up a glass to drink. Place a stand-up mirror on a table to do makeup, shave, or brush hair or teeth. All of these take the workload off the shoulder muscles.
- Modify tools with foam curlers or tubing to thicken the grip of forks, toothbrushes and other everyday tools.



The Arm Thing, being used by Ron Edwards, co-inventor with his wife, Linda

- Showers instead of baths minimize falls. Use a bath bench or seat while showering. Rest the elbow on the shower wall to wash hair.
- Getting up from a toilet seat is easier and safer with a higher seat. Use a raised toilet seat or a bedside commode positioned over the toilet.
- Prop books or magazines on a table, bookstand or music stand. The eraser end of a pencil can serve as an inexpensive page-turner.
- For working at a desk, rest forearms and elbows on chair arms or desk to relieve stress on the upper arms. A wraparound desk is especially helpful as it supports the elbows without reaching. A keyboard wrist rest can help to support hands, and on-screen keyboards minimize stress on the wrists at the computer.

For more information:

See “Accessibility at home,” page 32; Dressing,” page 37; and “Swollen extremities,” page 59.

“Telephone Access for People with Hand & Arm Weakness,” MDA/ALS Newsmagazine, October 2006

“Arming You with Tips for Living With Arm Weakness,” MDA/ALS Newsmagazine, February 2006

Dynamic-Living
(888) 940-0605
dynamic-living.com

Hygiene

See “Bathing,” page 33; “Drooling,” page 37; “Itchy scalp,” page 47; “Skin,” page 56; “Toileting,” page 60.

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Independence

Independence fuels a “fighting spirit.” Help your loved one maintain the highest possible degree of independence, changing strategies as the disease progresses. Figuring out ways around the limitations of ALS is a creative game, in which caregiver and loved one are on the same team and every victory for independence is a blow against the disease. (Note: The use of assistive equipment like wheelchairs is a move *toward* independence, not *away* from it.)

- **Be patient.** Independence for a person with ALS can be slow and painstaking. Sometimes it seems faster and easier for caregivers to do it themselves. Breathe, relax and be grateful for whatever degree of ability the person has.
- **Be collaborative.** Remember that people with ALS are capable adults even if they can’t communicate clearly. Through whatever means possible, discuss choices, make joint decisions and defer to the loved one’s wishes in decisions regarding their medical care.
- **Ask if they want help before helping.** Don’t take over tasks that still can be performed if the person is given adaptive devices and time.
- **Let your loved one use your hands.** When a person with ALS needs help with something, it can be very frustrating to have a helper take over the task and do it their way. The end result is seldom what the person with ALS envisioned. Your loved one doesn’t need a caregiver’s brain to plan things, just a pair of willing hands. Suggestions are fine, but ignoring the person’s ideas is demeaning.
- **Help set up your loved one with a computer and Internet access.** Help locate and install adaptive technologies that enable computer use when movement is limited or absent. Computers provide entertainment and social interaction and enable the person with ALS to help with household chores such as paying bills, tracking down information, hiring services and grocery shopping.
- **Use adaptive devices and strategies.** Consult a physical or occupational therapist for suggestions. See “Hands,” page 45, for suggestions of some adaptive devices to assist with *activities of daily living* (ADLs).

“The first time he used his power wheelchair, I took him in our new converted monster van to Costco. All of a sudden I realized he was no longer with me and in a panic I ran through the store looking for him. Where was he? In the candy aisle, looking for a treat!

He felt such a sense of freedom that he could wander through a store alone, which made me feel better.”

“I wished we had taken everyone’s opinion and got Mom’s wheelchair earlier. It took over 15 months to get the chair and by the time we got it, she could no longer drive it on her own. I was so hoping she might enjoy the independence.”

Other adaptations include rearranging household objects or furniture and changing the way a task is done (i.e., sliding something rather than carrying it).

- **Don’t sacrifice safety for independence.** It can be dangerous to leave unattended a loved one with a history of choking or falling. Families have found several ways to monitor loved ones, such as cell phones, baby monitors, emergency response buttons, friendly neighbors, etc. (See “Safety,” page 55.)

Itchy scalp

Some people with ALS seem to experience greater-than-normal scalp itchiness. There are a variety of possible causes, including dryness, yeast or fungus overgrowth, and stress. Check with a doctor and experiment to find the right remedy. Some things that have worked for others:

- Use gentle hypoallergenic hair care products.
- Shampoo hair in lukewarm water — hot water strips away protective scalp oils. Towel dry gently, not vigorously. Don’t wash too frequently or too infrequently.
- Use a humidifier.

- Over-the-counter remedies include Benadryl, Absorbine Jr. (applied liberally to the scalp), shampoos containing ketoconazole (such as Nizoral) or coal tar (such as T-Gel). Scalpicin Anti-Itch Scalp Treatment is a spray that can be used several times a day.
- In some cases of chronic itching or skin sensitivity, a physician may prescribe an anti-epileptic drug such as carbamazepine (Tegretol) or gabapentin (Neurontin), or one of the tricyclic antidepressants.

For more information:

See "Skin," page 56.

Jaw clenching

An uncomfortable tightening or chattering of the jaw in response to cold, anxiety or pain may occur in ALS. Relaxing medications such as diazepam (Valium) may help. Also see "Pseudobulbar affect," page 54.

Laughing, Uncontrolled

See "Pseudobulbar affect," page 54.

Lifts

As ALS progresses, mechanical lifts can save caregivers' backs, necks and shoulders from injury. Lifts are useful in transfers, such as from bed to wheelchair, or from wheelchair to toilet or bath, and in getting someone up off the floor after a fall. (See *Everyday Life with ALS*, Chapter 7, for ways of moving the person before a lift is needed.)

Lifts are operated mechanically or with motors, and have

“Even with the narrow hospital bed, I have to stretch when turning and caring for [my husband], and as a result my knees have been injured by pressing against the bedrail. I'm very cautious now and use slippery plastic under his shoulder and buttocks and a lengthwise-folded towel as a strap to pull his bent lower knee when turning him. I also pad my knees against the bed with a pillow.**”**

a sling of sturdy material on which the person with ALS sits. There are several types of slings, geared for different uses. Besides freestanding lifts, some operate on ceiling tracks.

Sometimes people with ALS are leery of lifts, feeling insecure and vulnerable swinging in this new contraption. Practice on other family members to get a feel for it and to allow your loved one to see it in action. Once the mechanics have been mastered, this is a terrific piece of assistive equipment.

Motorized lift chairs and uplift seat cushions (with a spring-powered seat) can boost a person upright when arms are too weak to help push up. Be sure the chair is correct for the person's height. These aren't likely to be covered by insurance policies, while mechanical lifts usually are. Check your policy.

To go upstairs and downstairs, a chair lift can be installed on the home's major staircase.

For more information:

Check with insurance, Medicare or the MDA equipment program for information on obtaining a lift.

"Give Me a Lift: The Right Tools for the Transfer," Quest, March-April 2006

For permanent or portable lifts, stairlifts or lift chairs, see ads in Quest or go online.

Massage

Regular massage by a professional or a caregiver is physically and emotionally therapeutic for people with ALS, and also can help them sleep. Gentle massage is preferred to deep muscle massage. Caregivers also will find a massage for themselves is a great way to ease stress.

Medical care

It's wise to have a primary care physician for care that doesn't involve ALS, and to have this doctor consult with your ALS physician when necessary. Regular medical checkups, flu shots and pneumonia vaccinations are essential to preventing respiratory complications. Conditions such as diabetes, cancer, hypertension, Alzheimer's, heart disease, etc., may affect what medications or treatments a person can take. For example, a respiratory system weakened by ALS may make surgery more risky.

Keep your ALS doctor and other doctors informed about all of your loved one's medications and conditions. Knowing as much as possible about ALS will help a caregiver determine whether a symptom is related to the disease or has some other cause. When in doubt, call someone on the ALS health care team.

Medication

To date, only one medication has been developed specifically for ALS — **riluzole (Rilutek)**. When started early in the disease course, it may add at least a few months to life expectancy, possibly by interfering with glutamate, a carrier of signals in the nervous system which may be overactive in ALS. Sanofi-Aventis (sanofi-aventis.us), headquartered in Paris, now holds a patent on the medication, but at least one other company is working on a generic version.

A few people have found that Rilutek caused upset stomach or skin allergies. An ALS doctor can help decide whether the drug is indicated.

Rilutek can cost up to \$1,000 a month. Medicare and most insurance companies cover it, and the National Organization for Rare Disorders (NORD) (rarediseases.org) has a Rilutek patient assistance program that can help with the cost. MDA and your health care team can help you explore all sources of coverage.

A number of other drugs are prescribed in ALS to help with symptoms such as coughing, depression, pain, pseudobulbar affect, etc. Insurance often covers these.

Cholesterol-lowering drugs known as **statins** may have some role in neuromuscular disease, but that role isn't clear.

Some studies suggest that taking statins may lead to ALS symptoms; others that the drugs may have anti-inflammatory benefit in the disease. Until the information is further clarified, ALS patients with high cholesterol should carefully consult with their doctors to determine the best therapies.

MDA-funded scientists are developing and testing several potential ALS drugs. Find out which drugs are being tested, and which clinical trials are open to participants, at clinicaltrials.gov, and by keeping up with news at MDA's ALS Division website, als.mda.org.

For more information:

See "Prescription drugs and supplements," page 141.

Clinical Trials in ALS
clinicaltrials.gov

National Organization for Rare Disorders (NORD)
(800) 999-6673
(800) 459-7599
rarediseases.org
Rilutek patient assistance program

Neck

Neck muscle weakness in ALS makes it hard to turn or hold up the head, leading to decreased mobility and — if untreated — pain, especially when turning the head, raising and lowering into bed, or rolling over. Poor neck posture also can impair breathing, swallowing and communication. Effective management of neck weakness can prevent or treat pain and injury. The key is to find the best methods of supporting the head.

Be aware that neck weakness makes the person more vulnerable to injury during transfers and when riding in a vehicle. If the head suddenly flops down, back or to the side, muscles and ligaments in the neck can tear, or the cervical spinal cord or neck vertebrae can be seriously injured. For some, even a slight jerk is all it takes to tear muscles.

Encourage and assist neck-stretching exercises (after consulting a physical therapist).

Two simple exercises:

1. Slowly turn the head to each side as far as possible without pain, hold and return to the center.



2. Tilt the head sideways on each side, so the ear points down toward the shoulder; hold and return to center.

Use a lumbar roll or cushion behind the lower back to prevent slumping when sitting, and to prevent the head and shoulders from tipping forward. Pillows under each arm also promote upright head position when sitting.

Work with a physical therapist to investigate different types of soft collars, neck braces and head supports such as a band around the forehead that attaches to a headrest. Alternating the use of collars and head support systems helps reduce pressure points and skin breakdown. Thin-cushioned skin dressings (e.g., Duoderm) also protect the skin.

Ensure the person's bed pillow isn't too thick, as that can cause neck strain. Try placing a rolled towel under the back of the neck, coupled with a thin pillow for the head.

Nightmares

Bizarre dreams or nightmares, coupled with morning headaches and mental fuzziness, may indicate respiratory problems in ALS. See Chapter 3, "Respiratory Issues," for more information.

Pain

Although ALS doesn't directly cause pain, it leads to some painful secondary conditions. It's estimated about two-thirds of people with ALS experience chronic discomfort or pain, primarily due to muscle cramps, pressure sores, stiff joints, overstretched muscles and **spasticity** (jerky movements caused by rigid muscles). Smaller involuntary contractions (twitches) are called **fasciculations**.

There are several avenues to pain relief:

- **Rest and sleep.** Lack of sleep can make anyone especially sensitive to discomfort. A good night's sleep or a few days' rest from overexertion sometimes can relieve aching and cramps. See "Sleep," page 56, for ideas on more comfortable sleep.
- **Check equipment.** As the person becomes less mobile, pain can arise from ill-fitting equipment, such as incorrect wheelchair seating. Any prolonged position can contribute to pressure sores. See "Positioning," page 51; "Pressure sores," page 53; and "Sleep," page 56, for more.

Ensure your loved one always is positioned correctly in the bed, chair or wheelchair. Experiment with cushions, mattresses and pillows until the right support is found. If it's impossible to get comfortable, investigate a change in bed or wheelchair.

Some people with ALS become profoundly sensitive to anything touching them. A light blanket may feel like a heavy weight or an annoying sensation when the person can't move to get comfortable.

- **Relieve coughing.** Prolonged coughing from the flu or from weakened respiratory muscles can become exhausting and make the chest muscles sore and achy. See "Assisted coughing," page 67, to learn how to help relieve this kind of pain.
- **Movement and touch.** Stretching, exercise and light hand pressure can help reduce pain. *Every day*, encourage, assist or perform range-of-motion exercises — these are critical for preventing or easing pain. Range-of-motion, if not too strenuous, can feel like a gentle massage. (See "Contractures," page 36, and "Range-of-motion," page 54.)

Applying heat, such as microwaveable moist heat pads, directly to the area of discomfort can provide relief. Warm baths or showers may work.

Don't forget massage. Gently massage the painful area till it relaxes, or have the person sit facing the back of the chair or lie down for a relaxing back rub.

Doctors have recommended leaning over and pretending to stir a big pot as a way to work out cramps. Moving the arm as if crawling up a wall can ease frozen shoulder joints. A good stretch for the arms is pretending to pull up a zipper behind the back.

- **Medicate.** Sometimes it's necessary to consult a doctor about medications for spasticity and cramps. It can take a while to find the right medication and dosage that reduces spasticity without increasing weakness. Some common medications include Baclofen, Zanaflex, Neurontin, Lyrica, quinine (Qualaquin), magnesium and clonazepam (Klonopin). Arthritis medications or over-the-counter nonsteroidal anti-inflammatories (Ibuprofen, Aleve) sometimes work. In states where marinol (medical marijuana) is legal, it's been found to relieve some pain, accumulation of mucus and spasticity.
- In advanced cases, such as for those in hospice care, narcotic pain medication may be prescribed.

For more information:

"Marijuana as Wonder Drug," Boston Globe, March 1, 2007

Living With Chronic Pain: The Complete Health Guide, by Jennifer P. Schneider, M.D., Ph.D., Hatherleigh Press, 2009

Making Friends with Pain: Learning to Live Well with Chronic Illness, by Elizabeth Flora, Sadie Books, 1999

Mayo Clinic on Chronic Pain: Practical Advice for Leading a More Active Life, 2nd edition, by Jeffrey Rome, Mayo Clinic, 2002

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

“An adult who can be a rock during tough times doesn't have to be the parent. Our kids got close to our social worker and also had friends whose parents listened as well. Sometimes it's best if it is someone not right in the middle of the situation. **”**

Parenting

ALS can strike as early as the teens and as late as the 80s. It very often hits those in their prime years, who are raising children or planning families.

Men and women with ALS have conceived and borne children after diagnosis. Sexual organs aren't directly affected, though sexual activity may be influenced by immobility or discomfort (see "Intimacy and sex," page 121).

Mothers and fathers with ALS find many ways to relate to their children. Activities may change, but children's need for a parent's love and attention does not. For some parents with ALS, having a child provides a reason to keep living and fighting the disease.

For more information:

See "Pregnancy," page 52. "Children" and "Parenting" in Chapter 6 explore emotional and practical issues of parenting.

Positioning

Proper body alignment and support can forestall a variety of problems, like pressure sores and joint pain. As noted by one woman with ALS, "One hour spent with an arm unsupported is miserable and can result in days of shoulder pain and sleepless nights." In general, the head, shoulders, hips and feet should be aligned and not too flexed or overextended. Caregivers have several strategies to achieve proper positioning.

- Lots of pillows, in different sizes and firmness, stabilize a person in comfortable positions. When side-lying, put a pillow between the person's knees to keep hips in alignment. For back-lying, put a pillow under the knees and elevate the arms slightly

with pillows. Use more pillows to “float” heels above the bed and keep feet from flopping to the sides or downward (see “Feet,” page 45). Large body pillows, rolled towels and air cushions work for some.

- There’s no such thing as a one-size-fits-all wheelchair. Consult an OT or PT to customize a chair to your loved one’s unique body proportions. Important measurements include depth of the seat, position and height of the headrest, distance from seat to footrest, height of armrests, and backrest trunk support. Finding and fitting the proper wheelchair cushion is a crucial element.
- A wheelchair wraparound lap desk helps support the trunk, arms and hands.
- In bed, a drawsheet is a key positioning aid. The drawsheet is placed on top of the bottom sheet, extending from the person’s shoulder to buttocks with at least 6 inches of sheet remaining on each side. Grasping and pulling on the sheet (alone or with a helper) allows even a large person to be rolled to the side or hoisted higher up on the mattress. Ask an OT or PT for a demonstration.
- Silky sheets and pajamas can make positioning easier.
- No one should have to sit in the same position all day or lie in one spot all night. When your loved one no longer can change positions, an important part of caregiving is helping shift their limbs or entire body.
- If the person keeps sliding out of the wheelchair, try a seat belt or rubbery shelf liner on the seat and foot supports.

“What helps me is being in a wheelchair with armrests to support my arms and a lap tray across the armrests to put my hands on. The armrests are cupped which helps keep my elbows from falling off.

If I get too low in the chair the upward pressure of my arms in my shoulder joints gets painful, so I need to be pulled up. Some non-skid material on my cushion helps prevent sliding down so I don’t need to be pulled up often.”

“When we first had him, I had this fear that he would know that I was different and that he would not bond with me, but they’re just so adaptable and they love you for who you are. I guess I associated being a mother so much with the action, changing a diaper and so forth. It’s almost as if I’m half sister, half mother, because he and I are always being pulled around by Dad.”

We laugh together all the time. I sense that he does know that I’m his mom. So it really has been eye-opening and humbling to me to see the love there.

What I wasn’t really aware of was just the everyday miracle of having a child in the house. Instead of just the sound of the TV or silence, it’s so cute to hear him laughing and talking to himself. I find it wonderful medicine for me.”

For more information:

See “Pressure sores,” page 53; and “Sleep,” page 56.

Everyday Life with ALS, MDA — Chapter 7: Transfers

Pregnancy

With ALS there are many factors to consider when contemplating having a child. For women with ALS who are considering pregnancy, these factors should be taken into account:

- The enlarging uterus will push up against the diaphragm, squeezing the lungs, which can exacerbate weak breathing.
- If a Caesarean section is required, certain types of anesthesia may be a problem because of the risk to respiration.
- Vaginal delivery requires strong muscles in the lower body.
- ALS in some cases may be hereditary.
- Prolonged bed rest can lead to significant loss of muscle mass, which probably won’t be regained in ALS.

However, many women with ALS have given birth. A child may give both parents joy and hope that will strengthen them in the battle against ALS. Consulting many experts may help couples make this very personal decision.

For more information:

See "Parenting," page 51 and page 124.

"Having Children After an ALS Diagnosis," MDA/ALS Newsmagazine, July 2010

The Disabled Woman's Guide to Pregnancy and Birth, by Judith Rogers, Demos Medical Publishing, 2006

Pressure sores

Whenever skin is under prolonged pressure, tiny blood vessels are compressed, the supply of oxygen and water is interrupted, and skin starts to die. **Pressure sores (*decubiti*)** are caused by staying too long in one position and by medical equipment such as a ventilator mask or neck brace. Sores can lead to infection, long and inconvenient recovery periods, and in extreme cases, death.

Some cautions to observe:

- Nutritional or respiratory problems can cause skin to become very fragile.
- People who are immobile should have their skin checked thoroughly at least once a day. For light-colored skin, an area that stays red longer than 15 minutes after pressure is removed is the beginning of a pressure sore. For dark-colored skin, look for areas that are darker or purplish-blue in color.
- A developing pressure sore causes a burning, itching pain. Even if there is little or no redness at the spot, treat the area as a pressure sore by reducing pressure and cleaning the area. Prompt response to this early sign can make a huge difference in just a few days.
- Ensure your loved one always is properly positioned in bed, chair and wheelchair. Reposition at least every two hours or whenever asked by the person due to discomfort. See "Positioning," page 51.
- Protect body parts that get a lot of pressure (especially skin over bony areas like elbows, shoulders, heels, tailbone, shoulder blades, ears and the back of the

head) with pillows, pads, specially designed cushions or cushioned skin dressings such as Duoderm.

- When making a foam pad to sit on, don't cut out the center (as is done for corns or bunions) as this can further decrease circulation.
- When people no longer can reposition themselves in a chair or wheelchair, get a pressure relief cushion, like a ROHO or Jay cushion, as ordinary foam or air cushions aren't adequate to prevent pressure sores.
- Experiment to find the most comfortable and effective pressure reduction in bed, such as standard or alternating pressure air mattresses, real or synthetic sheepskin, foam mattress overlays and automatic turning mattresses.
- Beware of recliners, where people with ALS tend to spend a lot of time. Outfit the recliner with a pressure cushion or sheepskin padding; protect elbows and heels; and keep a close eye out for problems.
- When ventilator masks and neck supports cause skin breakdown, alternate with different masks or head support systems to give skin a rest.



“Dad just keeps on weeping all day. So much so that it's difficult for us to know if he is happy or sad. I am trying to do all that I can to make him a bit more comfy, but he just doesn't seem to like it. **”**

For more information:

See “Skin,” page 56; and “Sleep,” page 56.

Check out ads in Quest, and search the Internet for “wheelchair cushions” or “pressure relief cushions.”

Pseudobulbar affect (PBA)

Uncontrollable crying and laughing, out of proportion to the situation and sometimes out of the blue, is a symptom affecting 15 to 45 percent of people with ALS. Although not well understood, the problem (also called emotional lability or **involuntary emotional expression disorder**) seems to be related to degeneration of neurological pathways that modulate emotional expression. This degeneration appears to cause a “disconnect” between a person’s actual mood and his or her facial expressions.

PBA’s emotional outbursts cause some with ALS to shun social situations and to worry that they’re “losing it.”

Excessive yawning and jaw clenching may also be symptoms of PBA.

In October 2010, the U.S. Food and Drug Administration (FDA) approved the drug Nuedexta, made by Avanir Pharmaceuticals, specifically for the treatment of PBA in ALS and multiple sclerosis.

PBA is tough on caregivers. Excessive crying is very upsetting, while inappropriate laughing can feel insulting in some situations. It’s important for everyone to remember that this symptom isn’t related to true emotions, but to neurological “disconnects.”

- Is it PBA or depression? Check with a doctor for a questionnaire that rates episodes for frequency, duration, voluntary control and appropriateness.
- Medical treatments include Nuedexta, which is approved specifically to treat PBA in ALS; and antidepressants, which may work even though depression isn’t the issue.

Talk with your MDA clinic doctor.

- Caregivers should try to remain emotionally neutral during a PBA crying episode. Although the natural response is to comfort a crying person, that may actually make it harder to regain control. Change the subject or take a break.
- Sometimes people can shorten their PBA episodes by focusing on breathing patterns. Focus on breathing in for laughing and breathing out for crying.
- In social situations, briefly explain to others that this is something that happens with ALS, that it doesn’t represent true emotions, and that it will pass shortly. Don’t allow your loved one to become socially isolated due to this symptom.

For more information:

“FDA approves emotional-expression medication,” MDA/ALS Newsmagazine, January–February 2011

“PBA Symptoms No Laughing Matter,” MDA/ALS Newsmagazine, March 2006

Range-of-motion (ROM) exercises

Stretching and moving muscles and joints is **essential** to maximizing movement and minimizing pain. Blood clots, pressure sores, discomfort, sleeplessness and contractures are some of the potential consequences of immobility. Caregivers can learn ROM from a physical therapist. If a loved one still can perform exercises alone, encourage daily practice. When assistance is needed, caregivers should assist only to the point at which the person can do it alone. Once voluntary movement is gone, passive ROM



“Range-of-motion is extremely important. [My husband] got ROM twice daily with touching and massaging of hands and feet whenever we were with him. This was for our benefit as much as his, so we felt close to him. Our children were great about massaging their dad's hands and feet. **”**

should be performed by a caregiver every day.

For more information:

Instructions and diagrams for stretching and range-of-motion exercises can be found in *Everyday Life with ALS*, MDA — Chapter 9: Exercise Instructions.

Safety

In addition to being prepared for the unexpected (see “Emergency preparation,” page 39), there are some precautions to keep in mind for everyday safety.

Keep the house free of obstacles (see “Accessibility at home,” page 32). Remember that weak arms make it difficult or impossible to brace for a fall (see “Falling,” page 44), and know that knees abruptly can give way without warning. Be available to lend an arm during walking, and encourage the use of assistive equipment.

Using a wheelchair is much safer than attempting to walk when each step is a struggle. Some people with ALS resist the wheelchair but may be willing to use it when a lot of walking is required. Keep the seat belt on whenever the person is in the wheelchair — at home, in public, in the

“When I have to use the alarm button, it rings into the company. They answer quickly and ask, ‘What is your emergency?’ Well, most of the time it is my cat that has pushed the emergency button. No problem, the medic alert is there when I need it. There is even a reminder alarm for my medications. I never forget anymore. It is great! **”**

car, etc. A slight bump, a steep ramp or uneven terrain can make someone tumble out of the chair.

The caregiver and person with ALS also should carry cell phones with important and emergency numbers programmed in. A medic alert bracelet or medallion will alert strangers to call the doctor.

For more information:

See “Accessibility at home,” page 32; and “Emergency preparation,” page 39.

Everyday Life with ALS, MDA — Chapter 3: Home Modifications
“Keys to Safety,” MDA/ALS Newsmagazine, April 2006

Saliva management

See “Drooling,” page 37.

Shoulders

Immobility can cause a painful condition called *frozen shoulder*. Although the shoulder may move, it's stiff and movement causes pain. Range-of-motion exercises prevent this condition and are a component in its treatment.

Shoulder pain also occurs when the weight of the arms isn't supported, causing a constant pull on weakened muscles and the shoulder joints. Carrying heavy objects can cause the arm to be *subluxed* (dislocated). Ensure proper positioning by supporting arms with armrests or pillows. Caregivers also need to protect the loved one's shoulders during transfers, by not pulling on their arms to move them.

For more information:

See “Exercise,” page 42; “Hands,” page 45; “Positioning,” page 51.

Everyday Life with ALS, MDA — Chapter 7: Transfers; Chapter 8: Exercise; Chapter 9: Exercise Instructions

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Skin

Some skin changes have been noted in ALS, such as changes in the biochemical properties of collagen and elastin which run through the dermis, or middle layer of skin. Blood vessels in the dermis also display irregularities and protein deposits as ALS progresses. Poor nutrition and respiration can make skin more fragile.

Caregivers can take several steps to ease skin woes for loved ones with ALS.

For dry itchy skin:

- Moisturize the air with a humidifier.
- Hot water strips the skin of moisture; use lukewarm water for baths and showers. If a hot soak is a must, use bath oil.
- From the standpoint of cleanliness, it's usually not necessary to take a full bath every day; a "parts" cleanup with a sponge usually is sufficient.
- Use mild or soap-free cleansers. Avoid deodorant soap.
- Pat skin dry with a towel; don't rub.
- Immediately after patting dry, seal in moisture with the greasiest lotion tolerated.
- Shave with lotion instead of foam.
- Try over-the-counter itchy skin remedies, such as those containing cortisone. (See "Itchy scalp," page 47.)
- In some cases of chronic itching or skin sensitivity, a physician may prescribe an anti-epileptic drug such as carbamazepine (Tegretol) or gabapentin (Neurontin), or one of the tricyclic antidepressants.

For skin infections:

Fungal infections like "jock itch" can be caused by being seated all day, which creates warm, damp pockets in underarms, groin and skin folds. To combat infections:

- Dry skin completely after washing, using a hair dryer on the cool setting.
- Buy clothing that wicks moisture away from the skin.
- Use over-the-counter antifungal sprays, powders and creams; avoid cornstarch, talcum or other nonmedicated powders.

- Herbal/natural remedies include applying apple cider vinegar or vitamin E; eating six cloves of fresh garlic or six to nine garlic capsules a day; eating yogurt with live cultures; and taking more B-complex vitamins.

Healthy-skin nutrition includes:

- Drinking eight to 10 glasses of water a day; taking a daily multivitamin-mineral supplement; eating daily servings of foods rich in linoleic and essential fatty acids such as safflower oil, nuts, avocado, seeds, soybeans, salmon, tuna, shrimp and corn oils.
- Skin damaged by pressure sores requires extra protein, zinc and vitamins A, C and K. Vitamin C also builds collagen, which is adversely affected by ALS.

For more information:

"Protection and Prevention Are Keys to Comfortable Skin," MDA/ALS Newsmagazine, December 2002

Sleep

Helping the person with ALS sleep well also helps the caregiver sleep well. Taking extra time to ensure comfort when a loved one goes to bed can cut down on call-backs for repositioning later in the night. Some caregivers create a checklist of bedtime adjustments to make sure nothing is forgotten. Although comfort is an individual thing, common elements are pillows for stability and to prevent pressure sores, a blanket lifter to keep weight off the feet, and blankets that don't restrict weakened movements by being too heavy or tight (see "Feet," page 45).

A comfortable mattress is essential. Some people prefer "memory foam" mattresses or mattress toppers; others use air mattress toppers or invest in automatic turning or alternating pressure mattresses. Automatic hospital beds allow people who can operate a remote to reposition themselves; the height-adjustable feature protects the caregiver's back and makes caregiving a little easier.

“[My daughter] says the Mattress Genie 'is wonderful because it allows me to continue using my bed instead of replacing it with a hospital bed.' **”**

When sleep is difficult, over-the-counter or prescription sleep aids such as Tylenol PM, Ativan, Ambien, Lunesta or Unisom may be in order (check with your ALS doctor first). Other meds have drowsiness as a side effect, such as allergy medications like Benadryl or the herb valerian. Tolerance to these substances can develop, so they should only be used if necessary.

To deal with nighttime saliva, try elevating the head off the bed. A tricyclic antidepressant such as Elavil can both dry up secretions and cause drowsiness. Other suggestions can be found under "Drooling," page 37.

For more information:

"Sleep Aids: Low-Tech Strategies for Improving Sleep Comfort" MDA/ALS Newsmagazine, March 2007

"One Good Turn," Quest, September/October 2006

Sleep deprivation (for caregivers)

ALS caregivers may get up numerous times a night to reposition or help their loved ones, leading to chronic sleep deprivation. Often the problem isn't getting up, but the inability to fall back to sleep afterwards.

Researchers say chronic sleep deprivation can cause depression, fatigue, forgetfulness, lowered alertness, reduced creativity, inability to speak and write clearly, lowered resistance to disease, weight gain and increased risk of stroke, heart attack and adult-onset diabetes. Sleep-deprived people also are more likely to verbally and physically abuse their children, and are more prone to falling asleep while driving. It's a problem that needs to be solved quickly. (See "Caregiver emotions and stress," page 102.)

Strategies to get more rest while still providing nighttime care include:

- **Make it quick and quiet.** When getting up, don't turn on the lights (use a low-level nightlight if necessary), don't have a conversation or do anything mentally stimulating, and stay up the minimum amount of time necessary.
- **Don't try too hard.** If you can't fall back to sleep within 10 or 15 minutes, get up and do something relaxing, then return to bed as you feel yourself getting

drowsy. Performing a good all-over body stretch can add in relaxation.

- **Decrease caffeine, alcohol and nicotine.** Especially avoid caffeine in the afternoon, as it can contribute to sleeplessness at night. Although some people find that an alcoholic drink before bed helps them fall asleep, alcohol increases the likelihood of waking later in the night.
- **Power nap.** Aim for a short (15-to-30-minute) nap sometime during the "midday trough" between 1 p.m. and 3 p.m., when your body naturally wants to rest. A longer nap may leave you groggy and unable to sleep at night. If you can't fall asleep, just rest quietly with eyes closed for a brief period.
- **Go to bed.** A simple way to get more sleep is to go to bed a little earlier. It sounds obvious, but tired people often stay up to watch the late show rather than turn in earlier.
Record favorite late-night shows and try to go to bed and get up about the same time every day.
- **Check out your sleep space.** An uncomfortable mattress, snoring spouse, too much light, being too hot or too cold, pets who jump on and off the bed, and outside noises all are subtle distractions that make it hard to settle back down. Try eyeshades, putting pets elsewhere or buying a more comfortable mattress.
- **Three on, three off.** If possible, share nighttime caregiving duties. For optimum benefits, sleep experts recommend a three-nights-on, three-nights-off schedule, rather than switching with someone every other night.
- **Talk to your doctor.** Not all caregiver sleep problems are caused by stress or getting up in the night. Schedule an appointment if nothing else is working.

For more information:

"Give It a Rest: Tips for Sleep-Deprived Caregivers," MDA/ALS Newsmagazine, December 2001

Socializing

Isolation is a risk for those with ALS. Symptoms such as drooling, immobility, pseudobulbar affect or difficulty in communicating may make people reluctant to see friends or go out. But it's important to keep up social activity and be part of the world and the community, and it's possible to find ways to adapt in almost any circumstance. Socializing helps fight off depression and enables the person with ALS to make a contribution. It also can help relieve caregiver burnout when others can keep your loved one company. Continued involvement with the larger world makes ALS only a new part of life, not an end to old interests.

Sometimes friends are reluctant to stay in touch if the disease makes them uncomfortable. If friends or family members seem uncertain how to relate to the person with ALS, remind them he or she is still the same person, and encourage them to talk about things other than ALS. Your loved one will want to keep up the same interests as before — sports, politics, movies, etc. Friends can come over to watch a ball game or concert on TV as a way of simply being together.

Friends also can spell caregivers at times so they can take a break or get other things done. Make it clear that they aren't being asked to provide personal or medical care, but rather simply to be there to talk or call for help if there's an emergency. The MDA clinic or support group is a new source of friends who can share the ALS experience — for both patient and caregiver. Some develop a whole new family or community that will be deeply appreciated throughout the ALS journey.

Going to public events is feasible and enjoyable. Most public places are accessible to wheelchair users, thanks

“ My mom refused to be seen outside with her walker and as a result she gave up shopping, going to parties, and other fun things that she loved to do. I tried to talk her into using her walker to do things outside the home, but she refused. She lost a lot of good years that way. ”



to the Americans with Disabilities Act (see “Accessibility outside the home,” page 33). With an adapted vehicle or public transportation, people with ALS can continue to work, go to movies, ball games, kids’ activities, church, family events and restaurants.

For more information:

For more about social relationships, see “Friends” in Chapter 6.

The Company of Others: Stories of Belonging, by Sandra Shields and David Campion, PLAN Institute for Caring Citizenship, 2005

Making the Moments Count: Leisure Activities for Caregiving Relationships, by Joanne Ardolf Decker, Ph.D., Johns Hopkins University Press, 1997

Yes You Can!!! Go Beyond Physical Adversity and Live Life to Its Fullest, by Janis Dietz, Ph.D., Demos Medical Publishing, 2000

Back issues of MDA publications such as *Quest* and the *MDA/ALS Newsmagazine* can be found online at mda.org/publications, or by calling MDA Publications, (800-572-1717).

Spasms

See "Pain," page 50.

Stairlift

See "Lifts," page 48.

Swollen extremities

In some people with ALS, fingers, hands, arms, ankles, feet or legs may appear swollen, sometimes extremely so. This is *edema*, the retention of fluid, and it has several causes. Often medications used to control drooling cause the body to retain fluids, leading to edema.

Most commonly in ALS, edema is due to being immobile. Muscle activity helps push blood through the veins to the heart. Without it, blood pools in the veins, and the pressure causes water in the blood to leak out into surrounding tissue, causing swelling.

Edema can be very uncomfortable. While the person is in bed, elevating the swollen parts above the level of the heart can reduce edema; prop the hands, knees and feet on pillows. If using an adjustable hospital bed, note that it raises the knees, not the feet. It's necessary to put a pillow at the foot of the mattress to bring the feet up level with the knees.

When the person is out of bed, correct positioning is critical. Recliner-type chairs can contribute to swelling of feet and ankles because, with the footrest up, all the weight of the legs is on the calves, reducing circulation. Adding pillows in the gap between the chair and leg rest can help distribute the weight and improve circulation.

Whenever the person is sitting upright, allowing the legs to hang without support will cause severe swelling. Adjust

“My legs are ghastly. They look fine when I am lying down, but the minute I sit up they turn hideous shades of red and purple and blue and gray. If I am not careful to prevent as much swelling as I can, they are swollen like sausages by noon and miserably uncomfortable, and absolutely painful by evening.**”**

the length of wheelchair footrests or put a box or cushion under the feet to reduce pressure at the thigh and back of the knee.

Swelling of the hands can be minimized by using a lap tray to support the arms. The best lap trays (easily cut from plywood or plexiglass) surround the waist and extend back at the sides to support the elbows. Elbow pads will be needed, and the hands can be raised on a pillow.

Any exercise possible also will help, whether it's being assisted to walk a little, tightening and pushing with the calf muscles, or just range-of-motion exercises.

Wearing compression hose can help, and shoes that lace are far better than slippers at controlling edema. Keeping the legs cool also is more comfortable. Reducing salt intake sometimes is recommended, although it's important to keep up fluid intake.

Diuretics, which increase the production of urine, often are prescribed but should be used only if other methods aren't enough to prevent discomfort. Diuretics can counteract medications used to control drooling, can deplete fluids and require additional trips to the bathroom — all problems for people with ALS. The family and medical team need to figure out a balance between controlling edema and excess saliva.

Edema also can be a sign of congestive heart condition, cirrhosis or kidney problems, so be sure medical personnel check for these, especially if the eyes are puffy. Edema just in a leg can be a sign of a blood clot.

Teeth

Dental care for a person with ALS eventually will require caregiver assistance.

As it gets harder to hold a toothbrush or hold the mouth open, the first step is an electric toothbrush, sometimes with a Waterpik. A child's toothbrush or foam-tipped swabs such as Toothette Plus Oral Swabs with Mouth Refresh Solution can be used without water and are easier to get into a tight mouth. These are available at many pharmacies and medical supply stores. Biotene toothpaste foams less than others and is easier to swallow or spit out.

Some people with ALS bite their cheeks, lips and tongues. A strong mouthwash can help heal the sores. A variety of mouth guards, such as SleepRight night guards or plastic sports mouth guards, can be found in drugstores. A dentist can order a rubber bite block or a custom mouth guard. These guards also can hold the mouth open while teeth are brushed and be worn at night to prevent teeth grinding.

Massage may help relax a clenched jaw. Be careful not to force anything into the jaw, because the person may inadvertently bite down hard on a toothbrush or a finger.

Getting teeth professionally cleaned can be tricky when the person with ALS has excess saliva, a tight jaw, or trouble sitting up or lying back. Any serious dental work should be done as early as possible after the diagnosis to minimize these complications. In most communities there are dentists who specialize in helping people with disabilities; the regular dentist or county dental society may be able to recommend someone.

During a dental checkup or cleaning, strongly remind the hygienist to suction constantly and thoroughly to avoid choking on saliva. Sometimes caregivers take over the suctioning because they're more familiar with signs from the patient and have to suction the mouth frequently at home.

Medicare and some private health insurance may cover dental care. A prescription or letter from the ALS specialist can help ensure the coverage, though it's not a guarantee.

For more information:

For more on suctioning, see Chapter 3.

"Those Teeth Are Made for Brushing: Open Wide and Say Ahhh," Quest, December 2002

Disability and Oral Care, ed. by Dr. June Nunn, FDI World Dental Press, 2000

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications, (800-572-1717).

Temperature

Cold extremities — feet and hands — affect some people with ALS. If these problems persist, practical solutions such as extra socks and gloves, room heaters, massage, range-of-motion, hot water bottles or hot packs can help.

In hot temperatures, breathing can be affected. Extreme hot or cold environments can affect the functioning of ventilators (see Chapter 3).

Toileting

Naturally, people with ALS prefer to take care of their own bathroom needs for as long as possible. This can be a touchy subject and is best handled step by step as needs increase — and with humor and lack of embarrassment.

- **Remodel:** At home the bathroom should have a door wide enough for a wheelchair to enter, and enough space for it to turn around, i.e., a 5-foot radius. Removing the door and hanging a curtain instead may be all that's needed. Special hinges also enable wider door openings. Add grab bars around the tub and toilet.
- **Stay regular:** As managing the bathroom becomes more difficult, people are sometimes tempted to cut back on their liquid intake or "hold it in" as long as possible to minimize trips. This is a bad idea. Dehydration can lead to constipation (see "Constipation," page 35) and swallowing problems, which are uncomfortable and unhealthy. People with ALS have bladder and bowel control for the most part — they know when they need to go — (see "Urinary urgency," page 62), and always should go when they need to.
- **Raise the seat:** Even for those who can transfer onto and off the toilet independently, getting up from a seated position can be a problem. Many raised toilets are available, including seats to be added to an existing toilet, raising the height by 2 to 13 inches. Get the highest one that works because your loved one will need more height as ALS progresses. A new toilet with a very high seat can be installed; there are self-contained units that don't require drilling.

into plumbing. A less expensive option is a portable, adjustable-height bedside commode chair that can be placed over the toilet. Some commode/shower chairs have wheels, allowing transfers to take place in another room.

- **Lift with a sling:** A hygiene sling has a large cutout that makes toileting possible from the lift.
- **Say hey to the bidet:** Arm weakness may make it difficult to use toilet paper effectively (hand-held toilet paper extenders are available). One independence strategy is the bidet. Several models, including the Lubidet, Brondell or Toto Jasmine Washlet, can be installed in place of the seat on the existing toilet. They provide a gentle warm water wash and warm air drying. Some have remote controls that can be operated by hand or foot, or by someone outside the door. Bidets run from \$400 to \$800 and usually can be installed by the homeowner. Bidet wands are good for traveling or public restrooms.
- **Portable urinals:** Portable urinals can save several bathroom trips a day. A bottle-shaped urinal is a good solution for men. Portable urinals for women have larger openings and handles. These can be used independently or with a caregiver's help, in standing, lying or seated positions. It's best to put down a waterproof pad first.
- **Catheters:** Men can use external condom catheters; some experimenting may be necessary to find the right type. Foley catheters involve inserting a tube into the urethra and connecting to a drainage bag. These indwelling catheters are used by men and women, and don't interfere with movement or transfer. The bag has to be checked and emptied, and the insertion site needs to be cleaned daily. If the catheter becomes clogged, painful, leaky or shows signs of infection, the doctor may replace it. The drainage bag should be cleaned periodically with two parts vinegar or chlorine bleach and three parts water. A suprapubic catheter may be used when the Foley is too uncomfortable or causes continued infections. It's inserted directly into the bladder through the abdomen wall.

- **Cleansing:** The time may come when your loved one with ALS is unable to transfer to the toilet at all and must perform bladder and bowel movements while in bed. Adult diapers (which also may be worn under clothes when going out) or disposable bed pads under the person in bed are the usual solution.

Thorough, gentle cleaning is important after a bowel movement to protect against bedsores. Wearing rubber gloves, turn the person onto one side and wash gently with baby wipes and a very mild soap such as Johnson's Bedtime Bath. Dry carefully and use some Desitin or other powder. A mix of Desitin with zinc oxide is comfortable and protective. To clean sensitive areas in the shower, hang two enema bags, one containing liquid soap and warm water, and one with warm water. Use one to clean the area, then the other to rinse.

- **Public restrooms:** In public places, there's usually a toilet stall which is big enough for manipulating a wheelchair. Still, your loved one may need assistance to transfer to the toilet and adjust clothing; a bidet wand may be convenient to carry. If the caregiver is the opposite sex from the person with ALS, ensure privacy by asking a bystander to stand guard or announcing "man (or woman) in the room" whenever someone enters. Or carry along a sign saying "Handicapped — opposite sex using restroom," and attach it to the door. See "Dressing," page 39, for other techniques that will make using a public bathroom easier.

For more information:

"Splish Splash: Easier Ways to Get Clean," Quest, January-February 2008

Transfers

See *Everyday Life with ALS*, Chapter 7.

Travel

People with all kinds of disabilities travel the world. To get around in town, investigate local public transportation and services for those with disabilities, or look into buying/renting a van that can accommodate a wheelchair.

Airlines, hotels and tourist sites have accommodations. Definitions of “handicapped accessible” vary widely, so it’s best to call in advance and find out exactly what’s available.

Cruises are a particularly comfortable way for people with disabilities to travel.

Airlines allow transportation of wheelchairs, respirators and other equipment, but usually as luggage. Damage to power chairs is not uncommon. If you’re flying to a distant destination in the U.S., you may be able to get a loaner wheelchair through the MDA equipment program in the city you’re visiting. Talk with your local MDA office about this before the trip.

For more information:

See “To Boldly Go” columns in back issues of MDA’s Quest for accessible destinations. (Go to quest.mda.org and type To Boldly Go in the search box.)

“Oh the Places You Can Go,” Quest, January–February 2007

“Travel Tips From People on the Go,” MDA/ALS News magazine, April 2005

101 Accessible Vacations, by Candy B. Harrington, Demos Health, 2007



Barrier-Free Travel, 3rd edition, by Candy Harrington, Demos Medical Publishing, 2009

There Is Room at the Inn, by Candy B. Harrington, Demos Medical Publishing, 2006

Able to Travel
abletotravel.org

Society for Accessible Travel & Hospitality
sath.org

Urinary urgency

Incontinence isn’t a feature of ALS because in general the smooth muscle of the bladder and bowel aren’t affected by the disease. But other muscles and nerves are involved and they may be weakened, making it harder to tighten the sphincter. At times the bladder may have a spasm, causing a powerful urge to urinate. Some medications may lead to more frequent urination.

Urinary urgency may prove temporary. Ditropan (oxybutynin) is often prescribed to relax the bladder and lessen urinary symptoms — watch for side effects including headache, dry mouth, constipation or diarrhea. Urinary urgency also can be a sign of a urinary tract infection, prostate trouble, fibroids in the uterus, or other causes, so be sure to have a thorough medical checkup.

Frequent awakening at night, followed by a need to urinate, may be a sign of respiratory problems; check with a doctor.

Yawning

Excessive yawning in ALS doesn’t mean tiredness. It can occur in half or more ALS patients, especially those with the bulbar-onset form. Yawning also can be a side effect of some drugs such as the antidepressant Lexapro.

Some people find sucking on a hard candy between the cheek and gums or chewing gum can stop the yawning — but beware if choking is a problem. Medications such as Effexor, Ativan or Klonopin may be prescribed. This symptom may be part of pseudobulbar affect; see page 54.



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Chapter 3

Respiratory Issues



Just Breathe

Decisions about ventilation are some of the most important that an individual with ALS will need to make. Information can make these decisions a little easier.

Underventilation dangers

Breathing requires the action of muscles — especially the dome-shaped diaphragm (the primary breathing muscle) located below the lungs and the intercostals between the ribs — to move air in and out of the lungs. Muscles in the abdomen, chest wall and neck also are involved in breathing. ALS can weaken all of these muscles, to the point that air no longer can move in and out of the lungs effectively. In this state of underventilation, or hypoventilation, the blood can't exchange carbon dioxide (which is exhaled) for oxygen (which is inhaled).

The effects of underventilation can be wide-ranging and serious. Difficulties related to respiration, including pneumonia, choking, sleep-disordered breathing, shortage of oxygen to the brain and cessation of breathing, often contribute to death for people with ALS. But careful monitoring and a choice of interventions can manage respiratory health in order to avoid a dangerous crisis and prolong life, sometimes by many years.

Many ALS specialists recommend getting a breathing test shortly after diagnosis. This gives baseline measures against which later tests of breathing can be compared. As respiration needs more attention, the ALS physician will recommend regular visits to a *pulmonologist* and *respiratory therapist* (RT) for interventions to support breathing.

Pulmonary function tests usually involve breathing into a computerized machine through a mouthpiece. One of the most important functions being measured is *forced vital capacity* (FVC), or how much air a person can expel as fast as possible after taking a deep breath.

“It's so much easier to focus on the positives when breathing isn't a moment-to-moment battle! **”**



It's important to see a respiratory specialist who works with neuromuscular disorders. Pulmonologists and respiratory therapists usually deal with lung disorders such as asthma or chronic obstructive pulmonary disease; many are unfamiliar with ALS. You need the respiratory team to understand that the breathing problems in ALS aren't a result of lung disorders, but of muscle weakness. Treatment for problems with the lungs won't correctly address ALS needs and can be dangerous.

NOTE: Supplemental oxygen, via an oxygen tank, is not a solution to breathing problems in ALS and may even be harmful. Supplemental oxygen is for people with lung diseases. In ALS, what's needed is help moving air in and out of the lungs.

Signs of respiratory insufficiency

Watch for changes in breathing, which may develop very gradually. Apparent fatigue, depression or sleep problems often are associated with developing respiratory weakness.

Symptoms of respiratory muscle weakness and chronic underventilation include:

- general fatigue, drowsiness, lethargy
- sleep disturbances such as nightmares, night terrors, sleep apnea (interrupted breathing during sleep) or sudden awakening
- morning headaches
- daytime sleepiness

- confusion, disorientation, anxiety
- poor appetite, weight loss
- excessive yawning or hiccups
- more labored breathing, especially when lying down
- rapid, shallow respirations with increased heart rate
- weakened or softened voice; speaking in short phrases; inability to sing or shout
- difficulty coughing and clearing the throat (weakened abdominal and throat muscles also contribute to this)

Any of these symptoms, seen regularly, should be reported to the ALS physician.

NOTE: *People with ALS don't usually show the kind of heavy labored breathing associated with overexertion.*

Simple ways to minimize respiratory problems

- Most important is to avoid exposing your loved one to any type of cold, flu or virus. (A weaker respiratory system can't effectively fight off infections.) This can push weakening lungs to pneumonia and respiratory crisis. If the person's respiratory system has shown any weakness, avoid close contact with anyone who is sick or who has been exposed to someone with a contagious illness. It's also important that caregivers protect themselves from cold and flu — and no hugs and kisses with your loved one if you have a cold.
- Raise the head of the bed while the person is lying down, either awake or asleep. You may want to purchase or rent an adjustable hospital bed or see if one is available from the MDA equipment program.

“It is much more manageable than I ever imagined. The trach care and suctioning takes very little time in your overall day and the vent does all the rest of the work. The only challenge is managing all the supplies, so talk to others who are vented if you decide to go down that route and find a good supply company. **”**

- Place more fans in the house to keep air circulating and fresh.
- Have the person with ALS and primary caregivers get a flu shot and a pneumonia vaccination.
- Increase the intake of fluids, as long as the person doesn't have trouble swallowing them (see Chapter 5).
- Help your loved one perform respiratory exercises, *only if* prescribed by a doctor or respiratory therapist.
- Include respiratory equipment or supplies as part of your preparation for emergencies (see “Emergency preparation,” page 39).
- Ask about medications such as expectorants, which help people cough up respiratory secretions more easily. In some cases, a doctor may recommend bronchodilators, which open up the bronchial tree.
- Review with the pulmonary care team a list of medications being taken. Some drugs can have an adverse effect on respiratory function, and can be switched to others with fewer respiratory side effects.
- Consider inserting a feeding tube to reduce the danger of aspirating food into the lungs (see “Feeding tubes,” page 89).

Coughing

Weakened abdominal and throat muscles in ALS diminish the ability to cough.

When a person with ALS can't cough up mucus and inhaled particles, these can fall back into the lungs, where they can cause irritation and infection. Weakened swallowing muscles also make **aspirating** (inhaling) food and liquids into the lungs more likely, which can irritate the lungs and bronchial tree. This can lead to pneumonia or acute choking. (See “Choking,” page 88.)

If your loved one's ability to cough isn't sufficient to clear secretions from the throat, lungs and trachea, you can learn manual or mechanical **assisted coughing techniques**. When coughing is weakened, it's important for caregivers to provide daily help in keeping lungs clear.



Assisted coughing

The simplest method is to place your hands or arms over the person's stomach, just below the ribs and under the diaphragm, while he or she is lying down or sitting.

After they take one or two deep breaths, firmly push in and upward toward the ribs as they cough. This will force air from the lungs more quickly and help push any secretions out through the mouth. Have a tissue or portable suction device available to collect any mucus coughed up.

This is best done on an empty stomach. If the person with ALS gets nauseous or throws up during the process, try gentler pushing. An increase or other change in mucus may indicate a respiratory infection needing medical attention.

NOTE: Let a respiratory therapist demonstrate these techniques so you'll be sure to perform them correctly. If the person with ALS has any discomfort or bad reaction, consult the therapist or physician.

For mechanical assistance, the Cough-Assist from Resironics, also called an *insufflator-exsufflator*, is a very handy, effective device. A mask is put over the mouth, and the machine blows air into the lungs, then reverses the flow, simulating a cough.

“When my husband was vented, the RRT recommended that I purchase a video cart on rollers. It works great. It holds his vent, CoughAssist, humidifier, suction and has another shelf with four plastic drawers that hold supplies.”

After either a manual or mechanical assisted cough, you may need to use a portable suction device to remove secretions from the mouth through a small tube. You can rent or buy a suction device (similar to those dentists use) from a medical equipment vendor or borrow one from the MDA equipment program.

Doctors also may suggest these cough assist methods:

- **expectorants**—prescription medications that thin secretions, making it easier to cough and clear the secretions
- **breath stacking**—closing the throat after each breath taken in through a mouthpiece and then coughing
- **oscillation vests** or airway clearance systems to “shake up” mucus in the chest

Emergency! Respiratory crisis

Progressive breathing difficulties are part of the natural history of ALS. It's wise to plan ahead, talking with the doctor and each other about dealing with breathing problems as they arise.

A breathing crisis can arise very quickly. Call 911 immediately if your loved one is showing severe signs of under-ventilation such as struggling for breath or inability to breathe. (See “Emergency! Respiratory crisis,” page 67.)

A **bag valve mask** (BVM or Ambu bag) is a normal part of an emergency crew's resuscitation kit or a hospital crash cart to help someone who's not breathing. (This is what doctors on TV dramas are calling for when they say, “Bag him!”) Ambu bags serve essentially the same purpose as mouth-to-mouth resuscitation and are available online or through medical supply stores.

Emergency medical personnel probably will want to administer oxygen in response to signs of respiratory distress. This is fine only if combined with breathing sup-

port. A written explanation from your physician should help explain the situation to emergency personnel. The ALS health care team should be notified of the emergency, and the specialist's contact information offered to the emergency team.

Planning ahead for assisted ventilation will help you avoid having to make decisions in an atmosphere of panic. See "Discussing options together," below.

Short of full-blown respiratory crisis, other signs, such as continued shortness of breath, fever, malaise or low pulmonary function test readings, require immediate medical attention. These could mean an acute infection.

Assisted ventilation options

When FVC reaches a certain low level, usually near 50 percent, the respiratory team will introduce the idea of *assisted ventilation*. There are two primary types: *noninvasive*, in which air is delivered through a mask and no surgery is involved; and *invasive*, in which a tube is surgically inserted in the trachea and attached to a respirator. Assisted ventilation can help your loved one feel less fatigued and safer, by improving sleep and decreasing the potential for respiratory crises. It can rest the breathing muscles and increase energy, allowing for more activities and enjoyment of life.

Discussing options together

Deciding which, if any, assisted ventilation to use in ALS is one of the most important and most personal choices you and your loved one will have to make. Discuss this well in advance of time to act on the decision, and review all the factors involved. An emergency room is a

poor environment for making such a critical decision — and a respiratory crisis may decrease your options.

It's difficult to face these decisions when you're trying to focus on ways to maintain life as you know it and to hold onto hope for a long survival or research progress. An absolute decision doesn't have to be made in the first few months after diagnosis. But getting the issues out on the table and exploring the options is important. Keep the door open for a while. The options may look different as time passes.

First, do some research through sources like those suggested in "Resources," page 73. Talk with your health care team; speak with other families of people with ALS. Check your insurance policy, and see what coverage is offered for respiratory equipment, supplies and day-to-day care.

Don't expect a consensus of opinion from everyone you consult. Even physicians who are knowledgeable in this area have different opinions. No one can tell you what's best for your family.

It isn't unusual for those in the early stages of ALS to declare they don't want to live with mechanical interventions such as a feeding tube, wheelchair or tracheostomy. But after living with ALS for some time, and making adjustments as abilities decline, each of these devices often seems more acceptable. Those with young children or other joys in living, or with certain religious beliefs, may feel that life is worth sticking around for, even if they need machines to help them move, eat and breathe.

Others may choose to decline invasive ventilation, and opt for pain control as their breathing further weakens and stops.

Obviously, this decision is very much an individual one. It should have input from close family members, and be made on the basis of complete information and a time of reflection. Ultimately the choice is made by the person with ALS; his or her wishes can be expressed in a medical power of attorney or living will document (see Chapter 7).

“The effects of the vent on my health — oxygen levels that prevent headaches and give me energy, stamina, appetite, and pink cheeks — are equaled by improvements in my quality of life. My days are no longer dominated by shortness of breath, tiredness, and fear of the next episode of lung congestion. My world has re-expanded along with my lungs. **”**

Noninvasive ventilation

Most people with ALS who need breathing help begin with **noninvasive assisted ventilation** (NIV), and some people are able to continue with this approach for many years. At first, the machine only may be needed during sleep — restoring a good night's sleep for both of you! Later it may need to be available during some daytime hours.

The most common form of noninvasive ventilation is the bilevel positive pressure type, in which air is supplied through a facial mask (**interface**) or other device on a timed cycle. This involves a small, portable machine that can be placed at bedside. The well-known **BiPAP** (bilevel positive airway pressure) machine by Respiromics delivers air at two pressures — a higher pressure to help the person breathe in more, and a lower pressure so they can breathe out with little resistance.

There are other NIV choices, which the pulmonologist will explain in recommending the best one for the individual situation.

Interfaces

The interface is the mask or other device placed over the nose, mouth, or mouth and nose, so a person can use assisted ventilation.

Ideally the person with ALS should try several interface options to get used to the feeling and practice breathing with the device in place. The RT should ask questions about sleep habits, nasal congestion and feelings of claustrophobia, and determine which interface might be most comfortable and supportive.

With patience, the user should get used to a comfortable interface in a few weeks or months. Begin by using it a few minutes at a time, gradually increasing duration.

A face mask is most common, but if users experience panic, claustrophobia or general discomfort at having something over the face, **nasal pillows** may be preferable. These fit inside the nostrils and put no weight on the face; some users can't even tell they have them on.

Some people use nasal pillows in the daytime and a face mask at night. Nasal pillows also make it easier to turn the head and wear eyeglasses.



Straps around the head hold the mask in place. If air escapes through the mouth at night, chin straps can keep the mouth closed.

Masks are designed to seal around the nose and/or mouth. When they leak, they can blow air into the face and eyes, and decrease the effectiveness of ventilation. Try different masks until finding one that seals properly. Gel- and air-cushioned masks are becoming more common, are lighter on the face and seal better. Those with bulbar symptoms may have the most trouble keeping the mask sealed. Some find the best solution is a custom-fitted mask or nasal plug made through the hospital's respiratory therapy department.

Some masks irritate the skin on the bridge of the nose or around the mouth. One solution is tucking moleskin, cotton or soft felt just under the nose part, being careful not to break the seal. Carefully clean and dress any sores or wounds. Apply an antibiotic ointment such as Neosporin while the respirator isn't in use.

All parts of an NIV device require regular cleaning and sterilization, including the tubing, filters and mask. Filters need regular replacement, and an enzyme cleaner may be required for the mask.

Settings

For NIV to be as comfortable and effective as possible, settings for air volume and pressure have to be correct and supportive. If they aren't, breathing distress will continue. Users have complained that their cheeks are puffed out or air bloats the stomach. These discomforts result from an air pressure or volume that's too high.

Usually your respiratory team, including representatives of the device manufacturer, will set the pressure levels based on a sleep study, or begin at low levels and adjust them with frequent monitoring. They can be increased gradually to the level your pulmonologist considers most supportive and your loved one can tolerate.

Setting up the equipment, interfaces and follow-up requires experience and careful adjustment to make it work properly for each person, although some machines come with software or controls and information that helps adjust them correctly. The respiratory team or the vendor supplying the equipment should be available to make adjustments as needed, or to perform overnight oximetry at your home. Try to find a support person with knowledge of ALS or neuromuscular diseases.

Oximetry is a measure of oxygen saturation that tells whether the respiratory device is providing enough support. A **pulse oximeter** should be used regularly to check oxygen saturation while using NIV. An RT can check the pulse oximetry on visits, or you can buy one for about \$350-\$400.

NOTE: Respiratory function is too important for guess-work. It's OK to make small NIV adjustments at home, but don't hesitate to call experts whenever you have concerns about the equipment, no matter how many visits it takes to get it right.

Several studies have suggested that NIV can actually slow the decline in respiratory function and make a significant difference in survival time in people with ALS. This applies to those with moderate or no weakness of the mouth and throat muscles. Those with more severe bulbar (mouth and throat) muscle involvement may be less likely to tolerate NIV and may have to consider invasive ventilation sooner in the disease course.

People with ALS and physicians hold strong opinions about the long-term use of NIV — for and against.

Whether your loved one decides to use NIV or invasive ventilation, close monitoring by respiratory experts is vital.

Invasive ventilation

Why would anyone choose surgery and invasive ventilation over NIV? Here are some reasons:

- It can take months to find the right mask or device and get used to NIV.
- Facial features such as a crooked nose or a deviated septum can make finding a mask that doesn't leak or breathing entirely through the nose difficult.
- NIV may aggravate sinus problems or cause severe abdominal distention.
- Some find anything on the face claustrophobic.
- Facial, mouth and throat weakness can reduce necessary jaw closure and ability to use a mouthpiece with NIV.
- Bulbar muscles are weakened so that the person can't speak or swallow or keep saliva out of the airway.
- The hours the person needs to use NIV increase from overnight to most of the day. Going out becomes more difficult if the person resists wearing the mask in public.
- Because pressure-based NIV can only *assist* breathing, as respiratory capacity deteriorates, the user slides back into the fatigue, poor appetite and anxiety of pre-NIV days.
- Pneumonia or a simple chest cold result in a respiratory crisis. As lungs become congested, NIV reaches its pressure limit more quickly, and less air is delivered.
- The person with ALS has problems swallowing and often aspirates food or saliva into the lungs, creating more respiratory distress.
- Invasive ventilation can allow the longest possible survival with ALS.

The most permanent type of ventilation is the positive-pressure ventilator with a surgically created **tracheostomy**. A ventilator is attached by a breathing hose to the tracheostomy tube, delivering air through the neck into the trachea (windpipe) on a timed cycle.

“I really wish [my husband] would have let me check more into how people can live with a vent and stay home with their family. He said he was never going to live that way. I sure wish I had pushed it more and showed him how others were living. Then just maybe this year he would have been here to see his son get his high school diploma, start college and see how well his other son enjoys singing music. How very proud he would have been of them. **”**

Tracheostomy surgery is considered minor and often can be done under local anesthesia with sedation. It's usually followed by several days or weeks of rehabilitation during which caregivers can learn how to clean and maintain the tracheostomy tube, change supplies and perform suctioning. Ask nurses and respiratory therapists all the questions you can think of during this phase.

Quality of life

At one time, people using invasive ventilation were kept in institutions because of their machinery. Nowadays, many people on total ventilatory support continue working, traveling, socializing and enjoying life. As with many of the changes brought by ALS, once the difficult decisions are made, the adjustment may prove relatively easy and well worth the effort. With the face now free of the NIV interface, some people are far more sociable.

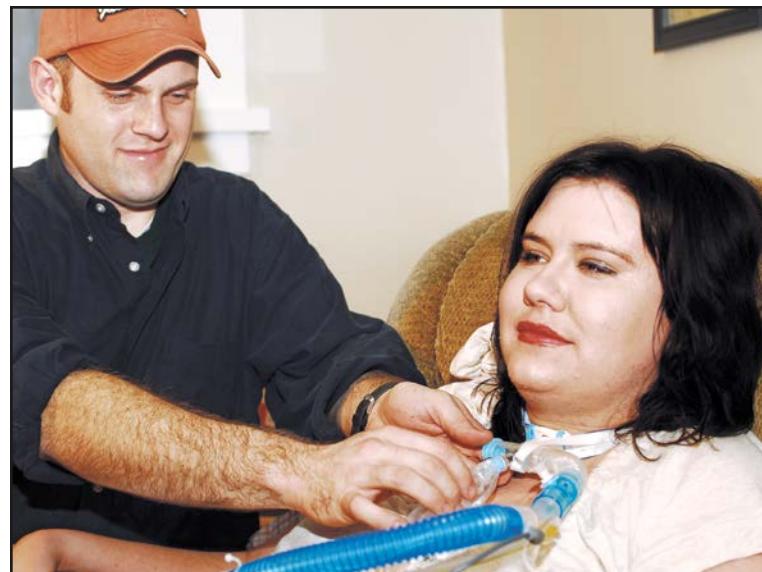
Today's vents are small, portable, relatively quiet and easily can be carried on a wheelchair. For those who can still speak, a speaking valve often can be added to the inflatable cuff at the end of the tracheostomy tube; this allows air to travel to the vocal cords, enabling speech. (If it's necessary for the cuff to be inflated all the time, the person won't be able to speak.)

In addition, a tracheostomy can provide a great feeling of safety. Permanent vents have alarms to alert caregivers to congestion or a disconnected tube. And invasive ventilation can return some energy as it relieves the exhaustion of poor sleep, prolonged coughing and labored breathing.

Problem solving

However, maintenance of invasive ventilation does require greater effort by the caregiver. Problems that arise may not be easy to fix without help.

- Whenever the trach or vent tubing is moved, it sets off an aggravating but not painful coughing spell. Vent hoses may pull on the tracheal opening, causing a sore throat-type ache. These problems are usually relieved by repositioning the hoses; a caregiver may require help from an RT or equipment provider to make this adjustment.
- Discomfort of the skin around the trach indicates irritation or infection and usually is easily treated with ointment.
- The most annoying discomfort is the same as with an NIV mask — air leaks. As with a mask, adjusting the hose position may help, but persistent problems may require a medical procedure to increase the size of the trach.
- Trachs carry a risk of incision infection, and increased respiratory infections are reported. At the same time, the trach is easier than NIV for clearing lung secretions and controlling respiratory status.



- Some people are prone to *granulation tissue*, an excessive growth of new tissue stimulated by the trach incision and presence of the trach tube. This tissue is delicate and bleeds easily and may make trach changes difficult. Granulation tissue can be handled with cortisone, silver nitrate and, in some cases, laser removal. Seek medical advice.
- Trach users need a backup ventilator.
- The battery operating the vent has time limits and will need to be charged regularly. An extra battery is necessary.
- If the person needs to go to a residential care home, some won't take vent users (see page 166).
- Vents often have temperature limitations, so avoid excessive cold or heat.
- When the ventilator is installed, contact the local electrical and/or gas power company and explain that this is a device required for life. Request a form to protect against the power being turned off and have it signed by the ALS doctor. A generator or other backup power strategies are advisable.

Diaphragm stimulation (pacing)

In 2011, the U.S. Food and Drug Administration (FDA) approved as a “humanitarian use device” a new method of respiratory assistance for people with ALS-caused breathing difficulties but with adequate preservation of the diaphragm muscle and the phrenic nerves, which stimulate it.

A humanitarian use device is one that the FDA has determined does not pose an unreasonable risk of illness or injury and for which the probable benefit to health outweighs the risk of illness or injury from its use.

Diaphragm stimulation, or pacing, involves a rhythmic stimulation of the major breathing muscle through electrodes, which must be surgically implanted into the diaphragm. The electrodes send signals to the diaphragm muscle, causing it to contract and assist breathing efforts.

The NeuRx Diaphragm Pacing System (DPS), by Synapse Biomedical, can be used in conjunction with nonin-

vasive ventilation (e.g., BiPAP) or by itself. The DPS does not slow or stop the progression of ALS, but it may delay the need for a tracheostomy, and may improve sleep and quality of life. MDA is supporting further studies of the device's effectiveness in ALS. For more, see Appendix D, “A Closer Look: Diaphragm Pacing System.”

What invasive ventilation means for caregivers

Those caring for people using invasive ventilation move into a new level of caregiving. A person with a trach must have someone available at all times to suction saliva and mucus plugs and respond to emergencies, such as a detached tube or power failure. However, nurses aren't required. Anyone, even an older child, can be taught how to suction and handle vent alarms.

Caregivers must decide who should be given the responsibility of ventilator care. For those hiring outside help, be aware that many home health care agencies only will provide registered nurses for people with trachs (see Chapter 8). This introduces a cost of several thousand dollars a month.

In addition, new supplies are required, including dressings, tubing, filters and batteries. See Chapter 7 to learn which costs may be covered by Medicare or private insurance.

It's possible for a person with a trach to shower but extreme care must be taken not to get water into the opening. Use a cloth or plastic wrap to keep water out.

Daily care for people with invasive ventilation includes:

- **Cleaning around the trach**, as part of bathing or washing up, using ordinary soap and clean washcloths. If there's a lot of mucus drainage around the tube, recurring infections or redness, more cleaning or use of ointments may be required.
- **Dressings** where the tube connects to the body must be changed.
- **The inner cannula**, a tube within the trach tube, must be cleaned daily, usually with dish soap or hydrogen peroxide. Some health care providers suggest changing the inner cannula every day.



- **Trach ties**, which hold the device and dressings around the neck, should be changed when they're damp or dirty.
- **The outer trach tube** needs to be changed regularly — some experts advise once a day, some say once a week. People prone to granulation tissue need to have the tube changed more frequently, as do those with respiratory infections. Changing the tube is usually a simple procedure.
- **Suctioning** is necessary to remove mucus or saliva that the person with ALS can't cough up. Some days people need suctioning two or three times, other days they require it a dozen or more times.

Generally suctioning can be done by a caregiver with a nonsterile disposable glove (not reused) and catheter (changed daily or several times a day). The respiratory care team will provide instructions. The caregiver passes a catheter attached to a suction device through the trach tube and down into the airway. This must be performed carefully because suctioning itself can push bacteria from the upper airway down into the deeper and normally sterile lung passages, adding to the risk of respiratory infections, including serious pneumonias. Be sure the gloved hand doesn't touch anything but the catheter.

You also can use an insufflator-exsufflator such as Respiration's CoughAssist (see page 67), to fit onto the trach tube and pull out secretions from both lungs at once. Use of the CoughAssist can reduce, if not eliminate, the need for suctioning.

Stopping ventilatory support

In a person with ALS who's using full-time invasive ventilation, muscles will continue to weaken to virtually total paralysis. If no alternative communication strategy has been learned (see Chapter 4), the person will be "locked in" with no way to move or communicate.

In this situation, or for other personal reasons, people with ALS sometimes wish to detach the ventilator and let nature take its course. It's important to discuss this possibility with the loved one so that his or her wishes are known before it becomes impossible to communicate. See "Advanced directives," page 148.

Ask the doctor about the options and consequences, and after a decision is made, be sure the doctor is aware of it. Putting choices in writing is a greater guarantee that your loved one's wishes will be followed; don't assume the health care team knows these wishes without being told.

Doctors can make the individuals with ALS completely comfortable and remove the ventilator. Medication is given to ease pain and respiratory distress.

Resources

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Respiratory issues

"Breathe Easy: Respiratory Care in Neuromuscular Disorders." MDA booklet (also available in Spanish)

"Breathe Easy: A Respiratory Guide for People Living With Neuromuscular Diseases." MDA video

“Breath of Life.” MDA video

Everyday Life with ALS, MDA — Chapter 5: Respiratory Issues

Coughing

Everyday Life with ALS, MDA — Chapter 5: Respiratory Issues

Respironics

(800) 345-6443

coughassist.respironics.com

Has free introduction and training CD-ROM and offers home care training.

Emergency! Respiratory crisis

International Ventilator Users Network

ventusers.org

Assisted ventilation options

health.groups.yahoo.com/group/NeckBreathers

Noninvasive ventilation

John Bach, M.D.

doctorbach.com/als.htm

“Treating Respiratory Problems in ALS: Patients Can Improve Quality of Life,” by E.A. Oppenheimer, M.D., Diana Guth, R.R.T., and Judith R. Fischer, M.S.L.S., alsindependence.com/Treating_Respiratory_Problems_in_ALS.htm

Interfaces

“NIV Masks: Finding the One That’s Just Right,” Quest, March-April 2008

Invasive ventilation

“Safe Harbor — Rediscovering Life on a Vent,” MDA/ALS Newsmagazine, July 2005

“A Tale of Two Vent Choices,” MDA/ALS Newsmagazine, August 2003

“A Breath of Fresh Air,” Quest, December 1998

“Home Ventilator Guide” and other publications

International Ventilator Users Network

(314) 534-0475

ventusers.org

Access Data

FDA database for reporting problems with vents and other medical equipment:

accessdata.fda.gov/scripts/cdrh/cfdocs/cfMAUDE/search.cfm

Quality of life

“Technically Speaking, It’s a Good Time to Have ALS,” MDA/ALS Newsmagazine, July 2007

“Safe Harbor — Rediscovering Life on a Vent,” MDA/ALS Newsmagazine, March 2006

“Life on the Vent: The Other Side of the Mountain,” Quest, June 2001

Problem solving

“What Will You Do If the Power Goes Out?,” MDA/ALS Newsmagazine, July-August 2007

International Ventilator Users Network

ventusers.org/vume/TreatingNeuroPatients.pdf

Diaphragm stimulation

“What is the NeuRx Diaphragm Pacing System (DPS)?” Synapse Biomedical, 2011

What assisted ventilation means for caregivers

“The Truth About Trachs,” Quest, July 2007

Tracheostomy Tube Adult Home Care Guide
tycohealth-ece.com/files/d0005/ty_ab3yzb.pdf

Chapter 4

Communication Issues



“ My granddaughters love to ‘make the machine talk.’ They are especially pleased when the ‘droid’ pays them a compliment, such as ‘pretty girl’ or ‘I love you!’ ”

“ I have typed the following message in a word document on the computer: HI: I HAVE ALS (LOU GEHRIG’S DISEASE) WHICH HAS AFFECTED MY SPEECH. I WILL TRY VERY HARD TO MAKE MYSELF UNDERSTOOD. ”

Stay in the Conversation

Losing the ability to speak is one of the most difficult challenges of ALS. (It should be noted that this doesn't occur for everyone with ALS.)

For the person with ALS, not being able to communicate is the ultimate isolation from the family, community or the world, especially if movement is also severely impaired. At some stage of the disease, thinking and expressing those thoughts may be virtually the only bit of independence left to someone with ALS.

For caregivers and other family members, never again hearing the sound of a loved one's voice and the absence of spontaneous conversation are key losses of family bonds and individual personality.

The loss of speech can change, and sometimes end, a relationship unless both parties make a great effort to learn new ways to communicate. Helping your loved one communicate benefits all involved and enables him or her to stay connected with others.

Thankfully, there are a number of devices — from low-tech letter boards to computers that mimic speech — that can keep your loved one in the conversation, no matter how much muscle is lost. (See "Resources" at the end of this chapter.)

Losing speech ability

The person with ALS may exhibit waning speaking ability (**dysarthria**) in several ways:

- The voice may become hoarse or raspy, or acquire a strained or tight quality.
- Words are slurred, or the person can't form words.
- The voice may sound nasal, as if "speaking through the nose."
- The voice may get fainter or softer; check respiratory status if this hasn't been done.
- Speech takes more energy and talking will be exhausting if respiratory health is declining.
- Problems in swallowing may be accompanied or followed by speech difficulty.

- The person will exhibit frustration, depression, anxiety or irritability at the inability to be understood.

Voice weakness or speech changes can occur at almost any stage of ALS, but tend to happen earlier in bulbar-onset cases.

Simple solutions

Here are some tips for being an effective speaking partner when speech is difficult to understand:

- Check for hearing loss and use an aid if needed.
- Give the person full attention and concentrate on the face.
- If you don't understand something, ask the person to say it again, slower or louder, or in different words.
- Ask the speaker to spell or change words that aren't clear.
- Ask the speaker to indicate when the topic is changing so you'll know the context.
- Repeat what you did understand so the person can simply fill in missing words.
- Talk in a relatively quiet place so that your loved one doesn't need to project as much, which can be fatiguing.
- If you notice that your loved one's speech is getting more slurred or the voice is getting weaker as you talk, encourage him or her to be quiet for a few minutes to give tired speech muscles time to rest.

Other assistance can expand communication.

At first, improvised sign language helps — pointing, gesturing, asking for clarification ("do you mean xxx?"), reducing conversation to yes or no questions, using a small bell to signal for attention. Make a simple handwritten letter board and point to letters or short words asking the person to say yes or no. Your loved one can scribble notes on a pad, magic slate or a dry erase paddle. Remember that markers are easier than pencils for weakened fingers, and encourage your loved one to write or spell key words instead of full sentences.

These methods are time-consuming but effective for a while at home. But it's important that the person with ALS be able to communicate with others besides the primary caregiver.

When speech begins to decline, a **speech-language pathologist** (SLP) or speech therapist referred by the MDA

clinic can help a person with ALS improve intelligibility and prevent fatigue. Techniques may include speaking slowly, taking replenishing breaths after a phrase or sentence, exaggerated enunciation or spelling out words when stuck. The SLP also will pay attention to swallowing and nutrition (see Chapter 5), which are affected by weakness in the same muscles needed for speech.

The SLP also may recommend a palatal lift, which is similar to a denture or retainer, if the main problem is a nasal voice due to weakness of the palate (roof of the mouth) muscles. Exercises to strengthen the muscles used in speech usually aren't recommended for ALS patients.

A voice amplifier can be of help if the major problem is a soft or weak voice. This will save the person energy from straining to be heard. If dexterity allows use of a cell phone or similar device, text messaging can improve communication efficiency.

You'll develop other speech shortcuts and substitutes, and may even have fun with them. One woman with ALS rang a cow bell to cheer at her grandson's baseball playoffs. At the next game, other parents and fans brought bells as well, and those rooting for the other team filled soda bottles with stones to shake.

After a tracheostomy (see Chapter 3), those who still have speech ability may be able to continue talking via a speaking valve added to the cuff at the end of the tracheostomy tube; this allows air to travel to the vocal cords, enabling speech.

A wireless paging system or baby monitor may be more appropriate than a bell for getting attention in the house. Paging can be part of an *environmental control unit* (ECU), which enables a person with ALS to control light switches, electrical appliances and more. For details, see *Everyday Life with ALS* or speak with a PT or OT.

Voice banking

Before speech declines too far, many people with ALS record their voices. This may include taping messages or stories for family members.

They also can record key sentences or phrases, names of important people, or other frequently used expressions on a Windows-based computer, saving the recording as .wav

files. The recordings can be transferred into some speech-generating *alternative augmentative communication* (AAC) devices. Save each message as an individual file to allow transfer to an AAC device.



Some AAC devices have programs that allow use of a pre-recorded voice. Audio files can be imported from your PC, or you can record your voice directly onto the device. Then the device will be able to "speak" in the individual's voice instead of a computerized voice.

Many people record themselves singing, laughing, saying signature expressions or reading stories for their children or grandchildren. Some record family memories or messages for loved ones. Many family members treasure their loved one's telephone answering machine messages simply because they have no other voice recording. Voice banking allows the family to keep much more. Everyone in the family will be gratified to hear their loved one's voice, and the person with ALS will be happy that he or she still can be heard. (See "Resources" at the end of this chapter.)

“Oh please record anything you can. How I wish I had spent more time recording my precious Daddy because I am still having problems remembering what he sounded like. I wish I had helped him record a journal. Anything you do for those who love you will always be treasured!”

Planning

When speech begins to deteriorate, the person with ALS and caregiver should start planning for eventual use of an AAC. There are many options, and a customized system may take three to six months to deliver, and several weeks to learn. Start looking at options and plan for the long term. Depending on the system chosen, these devices cost from several hundred dollars to \$14,000.

Check the MDA equipment program to see if there are devices to try. A university speech department or computer dealership also may have models to try without cost.

To obtain insurance or Medicare payment for the device, every feature has to be medically justified. Most funding sources require a speech pathology AAC evaluation, doctor's prescription and letter of medical necessity.

The SLP can help with AAC selection and preparation of paperwork needed.

Medicare will cover up to 80 percent of the cost for an AAC device. MDA assistance also is available for AAC repairs and modifications. Contact your local MDA office for details before making a commitment to an equipment supplier.

Medicare won't reimburse for the purchase of AAC devices that also feature functions like email, Internet access or word processing. Many AAC manufacturers whose machines include these capabilities also sell "dedicated" versions that prevent users from accessing anything but communication functions. Ask manufacturers and therapists about unlocking these features after you've paid for a device.

In some cases, a communication device may not be covered if the person with ALS already is in a nursing care facility or in hospice.

Role changes

All of the changes brought by ALS, including those in speech, require some adjustments in family roles. If your loved one with ALS has been the family spokesperson, the one who handled most family business or who maintained relationships, that responsibility will shift to others. Caregivers must try to accommodate the needs and wishes of all parties involved.

Speech difficulties can be very frustrating for the person with ALS and for caregivers. Patience is required as new means of communication are developed. A caregiver may have to convince the person that it's time to take steps to solve communication problems. As with use of a wheelchair or a ventilator, there may be some resistance to accepting this new level of loss. Offer all the support you can; patience, persuasion, the enticement of new gadgets, promises of help, a sense of humor and input from others who use AAC may help.

An inability to speak also affects the way a person is perceived by strangers, and even by friends and family. Some may assume the person can't hear or has a mental disability, and speak more slowly and loudly to "help." Caregivers can educate family and friends, but it's not necessary to try to teach everyone you encounter. Save the effort for those who matter.

A representative of a medical office or an agency such as Social Security may be reluctant to speak to anyone other than the patient/beneficiary. A power of attorney granting the primary caregiver permission to speak for the person can be faxed or emailed to the agency, and that usually solves the problem. Word these documents carefully to prevent hired caregivers from abusing the situation.

As communication techniques develop, it's important that the person with ALS continue to be "heard" and to participate in conversations, whether for business or pleasure. Some people with ALS carry a card such as this one:

Tips for communicating with me

1. *Find a position directly in front of me, at eye level.*
2. *Slow down. Our conversation will be at a slower pace than you're used to.*
3. *Please don't finish my sentences unless I ask you to do so.*

The primary caregiver can be an intermediary, gently reminding others to speak to the person directly, not to a companion, or to be patient with the technology or other means of communication.

With AAC devices, conversation will be slowed down. AAC allows users to prepare and bank some phrases and comments, but spontaneous remarks will have to be typed in.

Urge others not to talk over the person and to allow time for him or her to finish “speaking.”

Telephone

There are ways in which a person without a clear voice can speak on the telephone. The **TTY system** used by people with hearing difficulties is one; conversations are typed back and forth with text displayed on a visual screen or printed on paper.

For those with a strong voice but weak hands, speaker phones and some cell phones are designed to be used with voice input only. When the person can't operate a phone by either voice or hands, there's also **telephone relay** on the Internet, in which one person types his or her end of the conversation and an operator reads it out loud to the other party.

Your loved one also can use the AAC device to communicate on the phone. Some AAC devices even have telephone hook-up capabilities.

High-tech solutions

When notepads and speech therapy no longer help, high-tech solutions will be needed.

First, look in the help and support section on your computer and search for “accessibility.” A number of shortcuts are built into most computer systems to assist people with disabilities. Many relate to visual impairments, but there also are shortcuts and software such as **word prediction programs** that help speed up communication and save energy.



“ My dad refused to communicate with the Dynavox. It simply was too time-consuming for him. So I learned all the stories he wanted to tell, listening closely to what he was saying while he could talk. Toward the end I was blessed that sometimes if Daddy could even get a letter across to me I knew what he wanted to say. ”

“ Before the communication aid, I didn't answer the phone because no one understood what I was saying. With the machine, and the help of a good phone with a voice-enhancer, I can carry on conversations over the phone, which has made a big difference. ”

You also can add adaptive mice, special keyboards, large keys, and software that enhances accessibility and convenience to your existing computer, before you get an AAC device.

Higher-tech AACs include computers that speak as the person types, or input spoken words. If hand weakness makes typing impossible, there are “switches” (buttons) that allow computer operation with any working muscle, including the mouth, eyebrow, cheek and toes. (See **Everyday Life with ALS**, Chapter 6.)

Some AACs have on-screen keyboards that can be operated by a pointing device or the blink of an eye, and many have spell check functions or can be used as a complete computer. These devices can enable a person to continue working as well as communicate.

Mainstream devices such as smartphones or iPads often can run AAC applications (apps). Although much less expensive than high-tech AAC devices, popular hand-held devices don't always allow for alternative methods of operation, including eyegaze, head tracker or switch. Before buying, be sure the device still will be usable if hand ability is lost.

The best solution depends on the person's typing ability, functioning muscles, comfort with technology and the pur-

poses for the device. Do your research and select a device that's versatile and can be adapted as your loved one's functions and needs change.

Scientists are even testing *thought-controlled devices*, in which the user projects words from the brain into a computer with no mechanical interface. It's a long way from reality, but someday it could be just the thing for people with ALS.

The Right Words

by Ruth Anne Crowder

A communication board can be organized in categories such as Bodily Functions, Comfort, Diversion, Activity, or whatever makes sense for your lives. The person with ALS points to the correct choice or blinks eyes to indicate yes or no. Following are some phrases caregivers and their loved ones have found useful:

Bodily functions

Need to go to the bathroom

Bedpan

Urinal

Suction

Cough assist

BiPAP

Brush teeth, bathe

Change clothes or dress

I'm hungry

It's time to eat

Comfort

Too hot

Too cold

Turn

Move legs

Move arms

Move head

Scratch face

Scratch head

Scratch back

Scratch legs

Scratch arm

Raise legs

Lower legs

Blanket

Fan on/off

Raise/lower head

Diversion

TV on/off

Change TV channel

Music on/off

Change music

Radio on/off

Change radio station

Computer — bring/take away

Raise/lower window shade

I want to see out the window

Activity

I want to sit up

I want to lie down

I want to sit in a chair

Put me in/take me out of my wheelchair

Range-of-motion exercises

Take me outside/inside

I need the walker

Help me out of bed

Help me into bed

Physical assessment

Breathing problems

Pain

Fever

Feeding tube problems, trach/vent problems

(Reprinted from health.groups.yahoo.com/group/living-with-als)

Resources

Back issues of MDA publications, such as *Quest* and the *MDA/ALS Newsmagazine*, can be found online at mda.org/publications, or by calling MDA Publications (800)-572-1717.

Everyday Life with ALS, MDA — Chapter 6: Speech & Communication

Communications and Swallowing Solutions for the ALS/MND Community: A CINI Manual, by Marta S. Kazandjian, Singular Publishing Group, 1997

Losing speech ability

"The Strain of Silence: Coping with a Loved One's Speech Loss Calls for Inner Strength and Outside Support," *MDA/ALS Newsmagazine*, February 2008

"When Mouth and Throat Muscles Weaken," *MDA/ALS Newsletter*, February 2001

American Speech-Language-Hearing Association
(800) 638-8255
asha.org

“My family often refuses to ‘work with me’ to figure out what I am trying to communicate, choosing to throw up their hands immediately and say ‘I can’t understand you,’ and walk away or have me write it down. Then I feel like what I want to express is of no interest or importance to them, and I feel angry and of less value because I can’t speak intelligibly. It seems they are waiting for me to make it easy for them, rather than being willing to exert some effort to understand.”

“Not being able to express myself was like cutting off the essence of who I am.”

Simple solutions

Videos of people with ALS using low-tech communication
youtube.com/alscommunication

Laser pointer to use with communication board
55jer.com/communication/Laser_Pointer_Instructions.pdf

Voice banking

"Voice Banking & Speech Synthesis Technology," *MDA/ALS Newsmagazine*, July-August 2007

Planning

"Preparing for Silence: What to Do Before You Lose Your Voice," *MDA/ALS Newsmagazine*, March 2007

"Plan Ahead for Big Purchases," *MDA/ALS Newsmagazine*, January 2005

Role changes

"Partners Cope When Speech Is Lost," *MDA/ALS Newsmagazine*, October 2004

Telephone

"Telephone Access for People with Hand & Arm Weakness," *MDA/ALS Newsmagazine*, October 2006

AT&T relay

sprintip.com

Federal Communications Commission

(888) 225-5322

fcc.gov/cgb/dro/sts.html

Lists numbers to call in each state for speech-to-speech services.

Speech to Speech (STS)

(888) 877-5302 or (916) 448-5517

speechtospeech.org

Free national telephone relay service for people with poor speech.

High-tech solutions

"Microsoft Windows Vista: Speech-Recognition Feature Enhances Windows Experience," MDA/ALS Newsmagazine, April 2007

"People with ALS Share Personal Experiences With Life-Enhancing Devices," MDA/ALS Newsmagazine, February 2007

"Technically Speaking, It's a Good Time to Have ALS," MDA/ALS Newsmagazine, March 2006

"On-Screen Keyboards: Computer Access Made Easy," MDA/ALS Newsmagazine, February 2006

"Seeing Is Believing: Eye-Tracking Technology is an Option for Computer Users," MDA/ALS Newsmagazine, January 2006

"Little Communicators," MDA/ALS Newsmagazine, December 2005

"Assistive Technology is Still Emerging," MDA/ALS Newsmagazine, April 2005

"Free Dasher Programs Easy As ABC," MDA/ALS Newsmagazine, February-March 2005

"Naturally Speaking: Voice Synthesis Study Advances," MDA/ALS Newsmagazine, February 2004

Assistive Technologies: Expanding a Universe of Opportunities, by John M. Williams, Assistive Technology News and CTC Foundation

Computer and Web Resources for People with Disabilities, by Alliance for Technology Access, Hunter House Publishers, 2004

Smart Technology for Aging, Disability, and Independence, ed. by William C. Mann, Wiley, 2005

Speaking Up and Spelling It Out: Personal Essays on Augmentative and Alternative Communication, ed. by Melanie Fried-Oken & Hank A. Bersani Jr., Paul H. Brookes Publishing, 2000

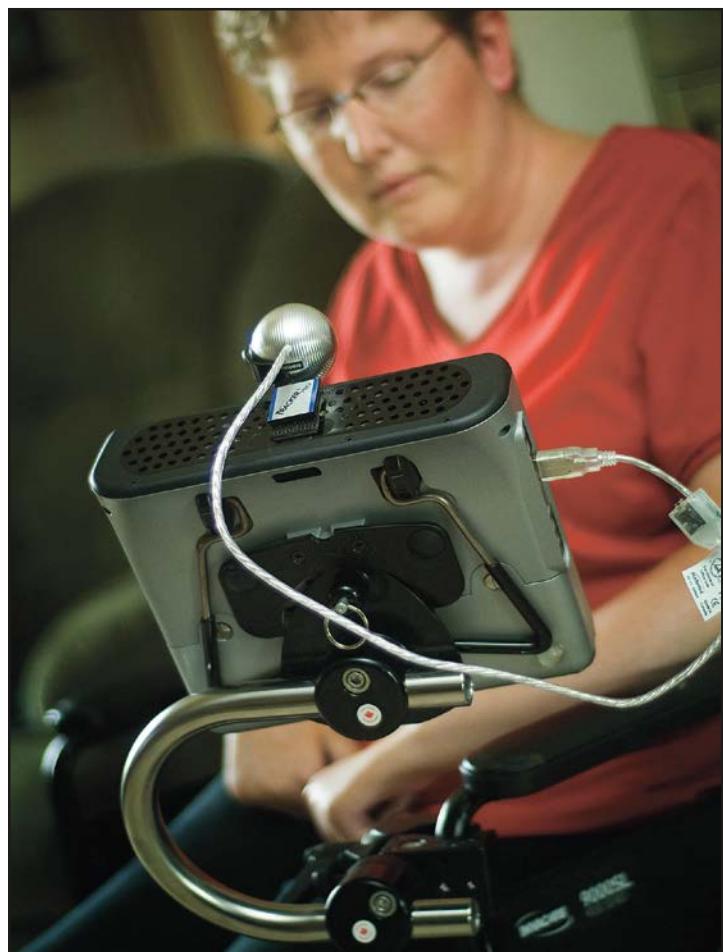
Microsoft Accessibility Resource Centers
microsoft.com/enable/centers/default.aspx.

Secondhand equipment
craigslist.org

"State of the Art Communication"
Film featuring people with ALS demonstrating several ways to use augmentative communication devices
als-communication.dk

United States Society for Augmentative and Alternative Communication
ussaac.org

Vocabulary for AAC users
aac.unl.edu/vocabulary.html



Chapter 5

Nutrition Issues



Maintaining Weight

It's a simple fact: Severe weight loss equals muscle loss. Therefore, it's important that a person with ALS not become underweight.

There are many reasons why people with ALS lose weight. Difficulty chewing and swallowing causes choking. Arm/hand weakness limits self-feeding. Other factors include decreased appetite, constipation, shortness of breath and nausea after eating, or fatigue due to the long and tiring process of eating.

Severe weight loss can be a red flag that a person's immune system is compromised, making him or her more susceptible to bacteria and other damaging intruders. Be sure to check this possibility with the doctor.

Studies suggest an improved survival with ***early, aggressive nutritional management***. While consuming adequate protein, vitamins and minerals is important for people with ALS, the most important dietary factor is the consumption of ***adequate calories***. This prevents deterioration due to poor nutrition. Frequent weighing is the best way to ensure caloric needs are being met.

Maintaining calories

At the same time as eating becomes more difficult, often breathing does too. This means the person has to expend enormous energy (burn more calories) just to breathe, making good nutrition even more important just as eating ability wanes.

At this point, the challenge for caregivers is to increase the amount of energy (calories) their loved ones consume without significantly increasing the amount of food they have to eat.

A ***dietitian*** or ***nutritionist*** generally can be recommended by your MDA clinic, and can advise on menu items that meet nutritional and caloric needs while still being easy to eat.

NOTE: Don't use diet foods, unless required due to other medical conditions such as high blood pressure or diabetes.

Healthy calorie boosters include adding nonfat dry milk or

egg whites to foods; using olive oil; and serving avocado, whole grain rice, pasta and 100 percent fruit juices.

Serving several small meals throughout the day can reduce the fatigue brought on by eating, as well as boost calorie intake.

One upside: Choices that are generally considered "unhealthy" are allowed for people with ALS, such as honey, butter, cream sauces, gravy, sugar and chocolate syrup, and frying rather than baking foods. These increase calories without increasing the amount that must be eaten, and should be acceptable as long as the person with ALS has no other medical condition restricting fats.

Instant breakfast powders, milkshakes and commercial nutritional supplements such as Ensure or Boost or their generic equivalents also are options. For higher calorie supplements choose the Ensure Plus or Boost Plus. Other high-calorie food ideas can be found by searching the Internet, asking at an MDA support group meeting or consulting the dietitian.

NOTE: Oral supplements with the word "plus" are better than the "high-protein" varieties, because they contain more calories and protein.

A well-balanced diet, with the recommended amounts of protein, fruits and vegetables, dairy products, grains and fats, will help the person feel as well as possible. Similarly, nutritional supplements may be recommended. Some sources promote specialized diets, such as gluten free, but there are no scientific studies and very few anecdotal reports that these have benefit in ALS.

Although there's no evidence-based information for the efficacy of ***supplements***, some physicians believe that creatine and antioxidants (vitamins C and E with other nutrients) may be valuable for people with ALS. Others advise a daily multivitamin. Use caution with fat-soluble vitamins (vitamin A, D, E, and K) as these are stored in the liver and can become toxic if taken in mega doses. To be safe, don't take more than 100 percent of the Recommended Dietary Allowance (RDA) of any supplement. Remember, supplements are that — they add to daily nutrition acquired from food, they don't substitute for it. Be wary of strong statements about nutritional supplements curing ALS.

Eating

Chewing and swallowing difficulties (*dysphagia*) make mealtimes exhausting for the person with ALS, due to the need to concentrate and go slowly to keep from choking. It's also a tiring and time-consuming process for the caregiver. Sometimes a person with ALS eats so slowly during a family meal that their meal turns cold and everybody else is finished eating and the dishes are cleaned up before the person is done — adding to a sense of isolation from the family.

A *speech language pathologist* (SLP) can advise on swallowing techniques to partially compensate for weakness and diet modification; the MDA clinic can refer you to this type of professional.

Other strategies include:

- Caregivers should cut up food into tiny bites before serving. Avoid dry and crumbly foods; these can be “glued” together with a soft substance like applesauce, gravy, yogurt, pudding, pureed baby foods or other soft foods.
- Liquids, soups and blenderized foods may be more easily sucked up through large-sized straws, such as those used for milkshakes at some fast food locations; they can be obtained at many discount stores. Other straws are designed for people with swallowing difficulties (see “Resources” at the end of this chapter).



Many ALS caregivers enjoy devising and sharing recipes for easily edible foods. You'll find many ideas on the Internet.

Weak arms and hands make holding and lifting utensils difficult, also inhibiting eating. There are many adaptive devices that help, such as lighter spoons with wider handles. An occupational therapist can provide more specific recommendations.

One useful device is The Arm Thing (shown in the photo at left), created by a man with ALS and his wife/caregiver, which enabled him to continue feeding himself by helping him lift a spoon or fork to his mouth. (See “Resources,” at end of this chapter.)

When it becomes hard to chew and swallow, *choking* and *aspiration* (inhaling food into the lungs) are concerns. (Sitting up for a while after a meal can help avoid reflux and possible aspiration.)

Now is the time for caregivers to be sure they know the Heimlich maneuver. See the section on choking on page 96 for more information.

A *feeding tube* (see page 89) can be placed while the person still is able to eat by mouth. This unobtrusive device allows the best of both worlds. Your loved one still can enjoy the taste of food and eat small portions with the family, while receiving the bulk of calories via the tube.

Drinking

Adequate fluid intake is essential for keeping saliva and mucus thin and avoiding constipation (see Chapter 2). Because drinking thin liquids can be difficult, and because drinking leads to urination, which can be time-consuming and require help, people with ALS sometimes don't drink enough.

Ensure that your loved one gets enough liquid — at least six to eight 8-ounce cups a day — to prevent *dehydration*, which can lead to physical deterioration, constipation, weakness, headache, feeling ill and thickened mucus that causes choking. Laxatives used to combat constipation also can contribute to dehydration.

Don't decrease liquids in order to decrease trips to the bathroom.

The first signs of dehydration are:

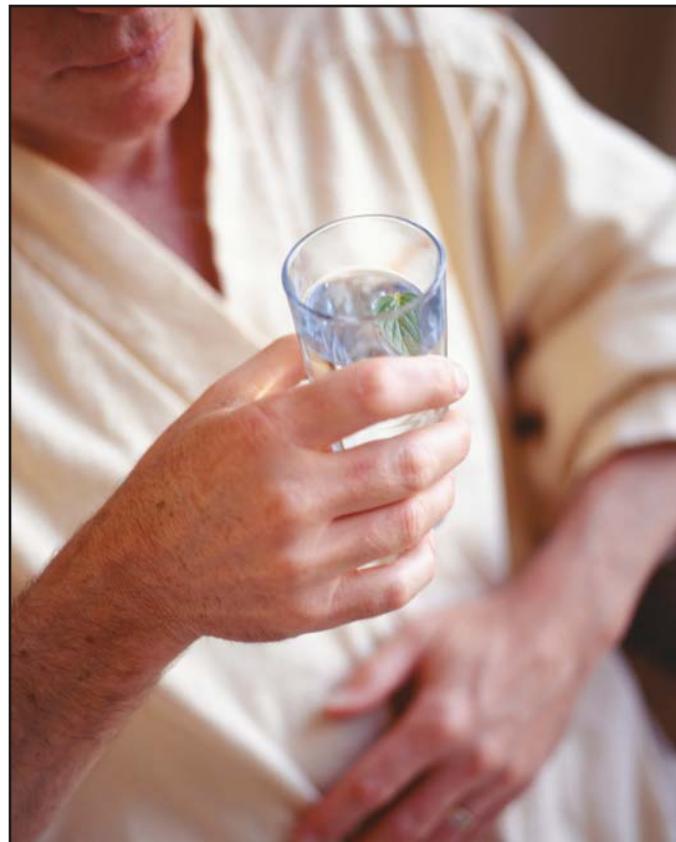
- thirst
- loss of appetite
- dry skin
- flushing
- dark-colored urine
- dry, sticky mouth
- fatigue
- chills
- decreased urine output — going for eight hours or more without urination
- few or no tears when crying
- increased weakness
- headache
- dizziness or lightheadedness especially from quick movement

Severe dehydration requires emergency help. The signs include:

- muscle spasms
- vomiting
- racing pulse and rapid heartbeat
- shriveled and wrinkled skin
- dim vision or sunken eyes
- painful urination, or little or no urination
- confusion and irritability
- difficulty breathing
- seizures
- chest and abdominal pain
- very dry mouth, skin and mucous membranes
- low blood pressure
- fever
- in the most serious cases, delirium or unconsciousness

“ A 2-foot length of water supply tubing became a straw that made up for his inability to raise a cup to his mouth. This is a great assist when traveling or drinking while watching television. ”

“ Mom was so dehydrated because she wouldn't drink or eat much to avoid the potty. ”



If thin liquids like water cause choking, serve thicker liquids like milkshakes, tomato juice, yogurt or fruit nectar, smoothies, gelatin and pureed soups. Powders like Thick-It and Thick 'N Easy add thickness without changing taste. Other thickening options, which also increase calorie intake, include baby rice cereal, mashed potato flakes, pureed baby foods, flour or cornstarch. Drinking liquids through a straw may help.

Alcohol, such as beer, wine or hard liquor, isn't harmful to people with ALS, but its effects may magnify existing problems with movement, coordination and respiration, increasing the danger of falling or choking. In a safe situa-

tion with other people around, and in the absence of other conditions that disallow alcohol consumption, the moderate use of alcohol is fine; drinks even can be put down a feeding tube. A few people with ALS experience muscle cramps when drinking alcohol. Also, check medications to see whether alcohol is restricted.

Most doctors advise doing what's pleasurable (e.g., enjoying coffee, soda, beer or chocolate ice cream). Just keep these in normal balance with nutritional needs.

Choking

As throat muscles weaken, the possibility of something "going down the wrong way" increases markedly and poses a real danger.

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

Choking is scary for both the person choking and the caregiver. Besides the suggestions above for making swallowing easier, there are several strategies for dealing with choking.

Clearing the clog

- Caregivers should remain calm and quickly assess the problem. Is the person able to get a breath, even a small one? **Don't hesitate** to call 911 if you can't quickly dislodge an obstruction that's blocking breathing. This is a life-or-death situation.
- Learn and practice the Heimlich maneuver and assisted cough technique (see page 67). Ask your MDA clinic for a demonstration. It's possible to use the Heimlich maneuver on a person who's in a wheelchair or unable to stand.
- Avoid back whacking. Focus on getting the obstruction **up and out**, not down the throat, because of the danger it will lodge more firmly or go into the lungs.
- An assisted cough technique, suction or cough machine can help clear a plug (see Chapter 3).

Avoiding choking episodes

People with even minimal swallowing problems never should eat while alone.

Caregivers can reduce choking episodes in several ways:

- Discourage your loved one from talking while eating, as this and other distractions make choking more likely.
- Remind your loved one to tilt the chin down while swallowing.
- Serve smaller but more frequent meals, avoiding dry, crumbly or large chunky foods.
- Encourage taking small bites and sips of liquids, and swallowing several times with each bite.
- Keep mucus and saliva thin by ensuring your loved is getting enough liquid and by using an over-the-counter expectorant (containing guaifenesin), or serving papaya, pineapple or lemon juice in water. Although there's no scientific evidence, many people find dairy products thicken their mucus.
- A nap before mealtime may give the person with ALS more energy for the tiring process of eating, thereby increasing safety.

Unexplained choking

One form of choking seems to come on out of the blue, even happening when the person isn't eating anything. A *laryngospasm* is a sudden tightening of the throat which occurs when liquid or saliva go the wrong way into the larynx; it also can be triggered by acid reflux, smoke, strong smells, emotion, alcohol, cold or rapid bursts of air, and even spicy foods.

In laryngospasm, the vocal folds go into a spasm that closes off the airway. The person may panic and be unable to breathe. After a few seconds, it's usually possible to suck in a thin, wheezing breath.

Laryngospasms usually clear before the person loses consciousness. There are different tricks for shortening their duration. Encourage the person to try:

- holding the head back and taking a quick "sniff" to break the breathing pattern;
- swallowing repetitively;

- exhaling in a gentle sustained breath, like a long soft whistle without puckering the lips;
- breathing in slowly through the nose rather than mouth; and
- pulling in breath from the belly rather than breathing shallowly.

Some people use Lorazepam Intensol — a concentrated liquid that must be kept refrigerated. Place 0.5 ml under the tongue or in the cheek, and let it be absorbed by the mucus membranes in the mouth.

During the spasm, caregivers should stay calm and offer reassurance, gentle back rubs and support. Although usually not dangerous, this experience is exhausting and upsetting for everyone.

Feeding tubes

If the person with ALS can't maintain weight, or eating and swallowing food or medication has become too exhausting, time-consuming, painful or dangerous, they should get a feeding tube. It's a much easier route to maintaining nutrition than trying to eat everything by mouth.

A feeding tube allows the person to eat what they're able, then supplements with adequate calories through the tube. Vitamins and medications also can be taken easily through the tube.

Early studies suggest a correlation between feeding tube use and longer survival and better quality of life.

People with ALS often resist feeding tubes because they feel it labels them an invalid, or signals they're giving in to the disease. One man said, "Getting the tube was an affirmation that I did have ALS, and that was one step closer to knowing that I was going to have to take additional steps to continue my life." But once they cross that psychological hurdle, the result is liberating and empowering.

When to get it

Doctors advise installing a feeding tube as soon as the person with ALS and family members are willing to have the procedure completed; those with bulbar-onset are especially encouraged to get a tube shortly after diagnosis. Even if the tube isn't used right away, it's in place when needed.



“There is so little that we have any control over with this disease. Fortunately, getting a feeding tube and keeping our nutrition up is one of those. **”**

As breathing becomes weaker, having a tube placed is more difficult because the person with ALS is more at risk for complications. Ideally the tube should be placed before the FVC (forced vital capacity, a respiratory measurement; see Chapter 3) falls below 50 percent of normal, as the procedure is safer and recovery is easier. And having the tube placed while the person still can eat by mouth allows for a gradual transition.

If FVC is adequate, the tube is inserted directly into the stomach via a **PEG** (percutaneous endoscopic gastrostomy) procedure. This may be done as a brief outpatient surgical procedure under local anesthesia, or your doctor may advise hospitalization.

“When we first started (doing the tube feedings) it was kind of scary to us, but now when we do it, it's just my mother and me and it's kind of a personal time. Sometimes we're completely silent, but it's a close, intimate kind of thing and I feel good about it. **”**

“My husband only lived 6 more months after getting his PEG, but it made life easier for all of us. It gave him more energy to interact with us. And I thank God every day that my children were spared the agony of watching their dad suffer the pains of dehydration. **”**

“Before the PEG, it would take him an hour to an hour-and-a-half to eat. I'd have to reheat the food several times and it was unpalatable. He got exhausted and gave up. It took an hour to get 200 calories in him. With the tube, I can get 200 calories into him in 10 minutes, and everything else he eats is for fun. **”**

— From “*Nutritional Lifeline: The Feeding Tube Decision, ALS,*” simplifiedtraining.com

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

Occasionally a tube is inserted into the small intestine, in which case it's called a **jejunostomy tube**.

Ensure your surgeon is familiar with the symptoms of progressive neuromuscular diseases and carefully discuss

all the options for placement and type/size of tube.

A few weeks after a tube is inserted and the **stoma**, or tube entrance site, has healed, a **Mic-Key button** may be attached. This skin-level, low-profile device, which resembles the valve on an inflatable beach ball, lies flat against the stomach, making it invisible through clothing and easy to sleep with. An outer tube is connected at mealtime, and nutrients are poured, dripped or infused through the tube from a syringe, bag or pump.

Quality of life

People who use feeding tubes for most of their nutrition report a much improved quality of life, and caregivers say that — after a few messy instances during the learning period — use and maintenance are easy. Time and energy are saved, allowing the family to focus on more interesting things. It gives control back to the person with ALS in terms of when to eat and how much.

On social occasions, family gatherings or at restaurants, your loved one can nibble on a bit of food or use the “taste-and-spit” method to savor a special treat. They may get a blenderized version of what everybody else is having, or they may receive their regular canned formula, then or later. Either way, they don't have to focus on struggling to eat, and can enjoy the social interaction.

The feeding tube won't protect against aspiration of saliva, but will lessen the likelihood of inhaling food or liquids into the lungs. Regular suctioning of the mouth can limit saliva aspiration.

What to feed

The tube can deliver blenderized or liquefied food made at home or a commercial formula. There are more than 100 commercial types, with Ensure and Jevity the best known. If a particular formula causes urgent bowel movements, constipation or an allergic reaction, a nutritionist can advise about alternatives.

Commercial formulas often are recommended over homemade formulas because they're nutritionally complete, sterile, and much easier and less time-consuming for caregivers than homemade formulas.

But some people want to eat “real food,” and actually can

get some of the food taste wafting up from their stomachs. If you chose to use a homemade formula, take care to prevent bacterial contamination. A dietitian can help ensure a balanced diet, proper food volume, adequate water intake and food safety procedures.

To make homemade formulas, an *industrial blender* is essential; standard commercial blenders simply can't blend food fine enough to go through the tube without adding a lot of water. An industrial blender like the Vita-Mix 5000 has a 2+ peak horsepower engine that can whirl food at 240 miles per hour. These blenders (which cost between \$300 and \$500) make it possible to blend up whatever the family is having for dinner without having to thin the mixture with so much liquid that it dilutes nutritional value. (See "Resources" at the end of the chapter.)

Home health nurses and MDA clinic staff can help the caregiver learn how to use the tube. It's important to deliver food at room temperature and as slowly as the person finds comfortable. Cold food or fast delivery can result in stomach cramps.

Tube feedings also can be given via a pump that hangs near the bed or wheelchair, very slowly delivering food over a long period of time.

Pills, including many tablets and capsules, can be pulverized, soaked and blended with liquid and put down the tube. A small mortar and pestle is handy for crushing pills. **Never crush or dissolve time-release or enteric-coated medications.** Check with your pharmacist to see if medications come in liquid form, and dilute them with four to five parts water to avoid stomach upset.

A heaping teaspoon of Metamucil powder every day can help prevent constipation (but take care not to clog the tube).

Maintenance

Clean the stoma regularly with soap; don't use hydrogen peroxide because it can lead to an excessive growth of granulation tissue that has the appearance of cauliflower. This excess tissue can be removed surgically or treated with applications of silver nitrate or cortisone cream.

Keep an eye on the stoma for leaking or irritation. These may be fixable by cleaning or applying ointment, but if they

persist call the doctor. Gauze squares can prevent skin irritation. Small amounts of bleeding and leakage of stomach fluids are common; increases may indicate that the tube is due to be replaced. Redness, pain, heat and wound drainage indicate infection, which requires medical attention.

Other feeding tube tips for caregivers:

- Flush, flush, flush with warm water before and after putting anything through the tube. Gently squeeze the tube as the water is running through to dislodge anything sticking to the inside.
- Flush the tube with water prior to giving medication and follow by flushing again.
- Tube feedings sometimes cause heartburn or nausea because the stomach is being filled too full, especially after weeks of undereating. Solutions include having your loved one stay upright for a period of time after tube feeding, feeding more slowly, giving smaller and more frequent feedings, trying different formulas and checking with a doctor about heartburn medications.
- It's time to change the tube when it's running slowly, can't be unplugged or is looking lumpy or deteriorating in some other way. If the stomach balloon that holds the tube in place deflates and pulls out, a replacement should be obtained within 12 hours, or it becomes more difficult to put in a new one.
- Black spots on the inside of the tube are due to Candida, a type of yeast. Digestive juices will kill any Candida that enters the stomach. Candida usually can be controlled but not eliminated short of a tube change.

Insurance

Some health insurance plans cover many of the supplies needed for assisted nutrition, including supplements and syringes for feeding tubes. To get maximum coverage, ask the doctor to provide a letter of medical necessity. See Chapter 7 for more information.

“ I don't even notice that I have the tube. I forget that it's there. ”

— From "Nutritional Lifeline: The Feeding Tube Decision, ALS," simplifiedtraining.com

Resources

Back issues of MDA publications such as *Quest* and the *MDA/ALS Newsmagazine* can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Maintaining weight

“Keep Calories Coming,” “It’s Still Eating,” “Equipment Corner (Gastrostomy Tubes),” and “Making the Switch,” *MDA/ALS Newsmagazine*, September 2006

Amyotrophic Lateral Sclerosis: A Guide for Patients and Families, by Hiroshi Mitsumoto and Theodore Mun-sat (Demos Publishing, 2001) contains extensive charts for maintaining optimal nutrition when chewing and swallowing are compromised.

The Dysphagia Cookbook: Great Tasting and Nutritious Recipes for People With Swallowing Difficulties, by Elayne Achilles, Ed.D., Cumberland House Publishing, 2004

I-Can’t-Chew Cookbook, by J. Randy Wilson, Hunter House Publishers, 2003

Meals for Easy Swallowing

Available online at mda.org/publications or by calling the MDA Publications Department at (800) 572-1717.

Eating

Coping Well With Home Enteral Nutrition
copingwell.com

Dysphagia Diet
(800) 633-3438
dysphagia-diet.com

The Arm Thing
MTE Devices
mtedevices.com

Oley Foundation
(800) 776-OLEY
oley.org

The foundation offers an equipment exchange program that provides free formula to those in need (shipping costs must be paid). To learn more about other supplies and equipment available in the equipment exchange, call (866) 454-7351.

Choking

Everyday Life with ALS, MDA — Chapter 5: Respiratory Issues, “Coughing and Clearing Secretions”

“All Choked Up,” *MDA/ALS Newsmagazine*, June 2005

Feeding tubes

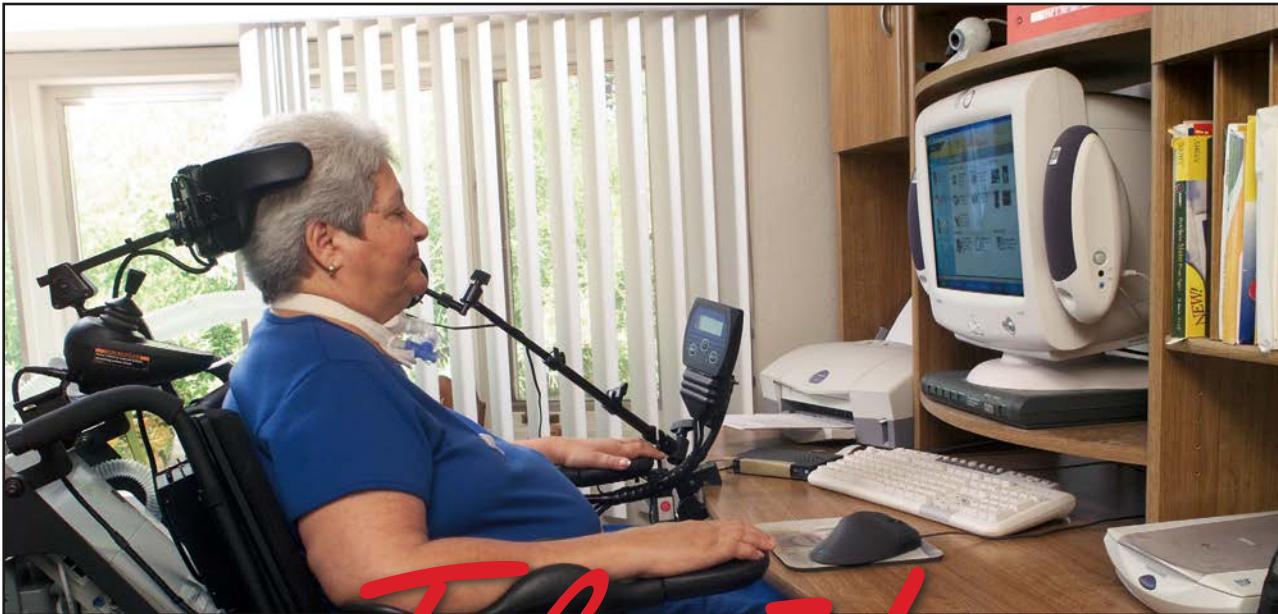
“Nutritional Lifeline: The Feeding Tube Decision/ALS” (DVD)

Simplified Training Solutions
(800) 344-6381
simplifiedtraining.com (free download of one chapter)

Homemade Blended Formula Handbook
by Marsha Dunn Klein, MEd, OTR/L and Suzanne Evans Morris, Ph.D., CCC-SLP, Mealtime Notions, 2007, mealtimenotions.com or (520) 323-3348. Although geared toward pediatric patients, the book contains extensive information about making nutritious blends for feeding tubes.

The Vita-Mix Corporation
(800) 848-2649
vitamix.com

A Medical Needs Program offers people with ALS factory-reconditioned industrial-strength blenders at a discount. Email household@vitamix.com for more details on how to qualify. Include the reference code 07-0036-0007.



Take 5!

Five Minutes Can Mean a Better Future

This is an exciting and challenging time in national health policy.

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Chapter 6

Emotions



Learning to Cope

A diagnosis of ALS unleashes powerful emotions for the person diagnosed and for close family members. On receiving the news, most people go through some version of the five steps in the grieving process — denial, anger, bargaining, depression and acceptance. Added to the mix is the fact that ALS is difficult to diagnose. Your emotions may have been on a roller coaster for months, but it's still a shock when the possibility of having the disease turns into the reality of a confirmed diagnosis.

The topics in this chapter cover some of the primary emotions, stressors and psychological issues that may occur during the ALS journey, along with ways to cope and sources of help.

(Note: If you're new to ALS, you may want to start with "Grief," page 115.)

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“ Give yourself a little time to process this information before you start making big changes in your own life. ”

“ [My husband] has had ALS now for three years. What helps him is knowing that he can't change it, but he can fight it. Not giving in or giving up is the key. I let him struggle until he asks for help. This is his way of knowing how much he is losing in strength. He can walk the dogs, or shall I say they walk him, but we have walkie-talkies to keep in touch. ”

“ No stage of my progression turned out to be the total nightmare I was dreading. Because of this, I decided long ago to live in the moment and not fret about the future. It took some practice, but this state of inner peace is well worth the effort. ”

“With decreasing physical abilities and increased discomfort, his behavior continues to worsen. He calls me names and says terrible things to me when I can't understand his speech or I don't do something exactly the way he wants it. At first, I thought it was just the frustration of his situation, but he is so different, I have to wonder. **”**

Abuse

It is important to recognize that caregiving is stressful. In an ideal world, caregivers provide love and care with tenderness and respect, and the loved ones receive each act of care with gratitude and consideration. But because nobody's perfect, impatience, resentment or other tensions at times may rise between them. For most caregivers and their loved ones, these feelings usually pass or are recognized and dealt with.

But when anger and contempt are out of control, the result may be physical or emotional abuse by either the caregiver or by the person with ALS.

A caregiver's abuse of the person with ALS can take many forms, including:

- sarcasm
- constant complaints
- noncommunication
- screaming, yelling or swearing at the loved one
- making the person ask several times for help
- poisoning the minds of other loved ones, including children, against the person with ALS
- blame
- accusations of self-pity
- ignoring requests or opinions
- shutting out family and friends
- bullying
- giving unprescribed tranquilizers or sleep-inducing drugs

- refusing to take the person to the doctor or follow doctor's instructions
- stealing money or valuables
- withholding medication
- rough handling
- physical assault

Needless to say, any type of abuse must be addressed immediately. Sometimes the caregiver needs a respite or a less demanding role, which can be provided by hiring a caregiver for several hours a week or creating a wider circle of people to share in providing care (see Chapter 8). Lack of sleep is a large contributor to abusive behavior; finding nighttime assistance may help a lot. Family or individual counseling is available through churches, hospitals and other sources. Many caregivers benefit from taking antidepressants or anti-anxiety medications.

If the caregiver cannot stop the abuse even after these solutions are explored, then it's better to leave than to continue the abuse. If no other solutions exist, it may be time to investigate assisted living or a nursing home.

Caregivers aren't the only potential abusers. Someone with ALS may direct his or her anger at the disease toward the nearest person — the primary caregiver. He or she may make outrageous demands, resist efforts to help, complain about the quality of care. Never hearing “thank you ... I love you ... I appreciate what you're doing for



Though ALS forced musician Richard Holland to give up playing instruments, he coped with his situation by using digital technology for composing his music.

“me” also can feel like emotional abuse. An abusive loved one adds sadness and pain to the caregiver’s role.

If the person with ALS continues this verbal and emotional abuse, be sure the MDA clinic has checked for dementia (see page 108) or other emotional changes related to the disease. Counseling, a wider circle of caregivers or medication may help. Unless it’s part of the disease process, having ALS is no excuse for rude or hurtful behavior, and caregivers should talk with their loved ones, explaining how they feel and describing behavior they would prefer. This conversation may open the way for a loved one to discuss their own emotions and needs.

Sometimes, when behavioral changes are part of the disease, the only choice is tolerance: A caregiver may love the person enough to ignore the abuse and not take it personally. Those who can’t tolerate the hurt aren’t any less noble — everybody has limited psychological resources, and nobody’s superhuman. It can help to become part of a support group and find other sources of care, even if only for a few hours.

For more information:

“Crimes Against People with Disabilities,” Quest, May-June 2008

“Rules for the Care and Treatment of Caregivers, MDA/ALS Newsmagazine,” February 2005

Acceptance and coping

Well-meaning friends may advise stressed-out caregivers to “stay positive.” That’s easy to say, not so easy to do. How a person learns to accept ALS, and cope with it emotionally and practically, depends on many factors, including the individual’s situation, personality, cultural background, resources and time demands.

People have different styles of coping with problems.

Some immediately jump in to find solutions; some remain indecisive; some deny or ignore a problem. Knowing your own and your loved one’s coping style will help both of you approach acceptance. Caregivers can build on the strengths of the existing patterns. For instance, for someone who wants to ignore the problem: “Let’s do something fun this weekend and try not to think about the diagnosis;

but let’s agree to start talking about it Monday.”

For people with ALS, acceptance may come gradually, in baby steps — “I have to take this medication … I’ll use this wheelchair but only at the mall.” Don’t try to shock the person into acceptance; let them go through it on their own terms.

Acceptance is important; without it people spend energy fighting reality instead of making the situation better. In our minds we know there’s no escape from ALS, but our emotions may need time accepting that truth. When grieving leads us toward acceptance, we can get on with the business of finding help, giving care and making the best life possible with ALS.

People with ALS get uncomfortable with being on the receiving end of help all the time, without the ability to give back. Caregivers can find ways for them to give back — helping with decisions, talking to children, bill paying online — and to reassure them that they’re giving something just by being around. Point out ways they still fulfill roles within the family.

As physical strength declines, it’s hard for people with ALS to accept the gradual inability to “do.” We’re a culture of “doing” rather than “being.” It takes tremendous effort on the part of caregivers to help people with ALS continue to “do” through equipment, help with daily activities and adjustments in activities.

It’s also important to help them learn to “be.” Many with ALS and their caregivers have found the experience an opportunity to develop spiritually and emotionally; to savor time with loved ones; to pray and meditate, revel in nature, enjoy learning and reading, explore their history and their thoughts; and to learn to live more in their minds and hearts as their bodies fail. As your loved one takes these gradual steps toward acceptance, stand aside and let him or her process the emotions and go inward. This will help him or her learn how to just “be.”

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

“This life is light years from the promising future ALS destroyed, but these are the cards in my hand. I can play or fold. There is no adventure in folding.**”**

“In taking care of my mom, I have to be the main decision maker. Sometimes she would just rather not deal with things. She's scared and worried so I'm learning to say 'This is what we are going to do, this is how we are handing this problem.' AND I'm learning to trust my instincts more.**”**

“As a caregiver I must guard against focusing on what this disease has done to him and try to focus on being the best caregiver I can be. I catch myself thinking about the 'skin and bones' look he has and it causes so much stress that I find I can't be the quality caregiver he needs.**”**

“I ended up in the hospital with a scare — I thought I was having a stroke — but it all checked out fine. I realized that I was running myself ragged and not getting enough sleep.

Even though everyone kept saying 'take care of yourself,' I just couldn't stop the cycle. I realized I needed more help.

I now have people that come in at night to help get [my husband] to bed during the week so I can get to bed at a decent hour. It took me a loooonnnnggg time to let go.**”**

One group of students studied ALS caregivers and found five types of coping styles:

- Taking it — being aware that they have no control over the disease and facing events as they occur
- Staying afloat — surviving, tolerating and attempting to maintain normalcy

- Losing ground — dwelling on loss and sadness
- Regrouping — regaining some control by getting assistance
- Holding together — making the most of the situation and adjusting to it.

Most people adopt some combination of these styles, surviving at times, finding new strength at other times, mourning losses as needed. Regrouping is probably the healthiest approach — it means adjusting to changes and finding help through equipment or other people. These styles help to modulate the more powerful emotions — not suppressing or denying them, but not letting them overwhelm either. Modulation can enable coping.

Drinking, overeating, escaping through television, dependence on drugs and the like aren't good coping measures. They'll exacerbate health problems and interfere with true rest, only serving to disguise feelings and frustrations. Don't fall into these traps.

There are some techniques that can help ALS caregivers and their loved ones accept and cope with what the disease is doing.

- **Exercise** can help caregivers work out some stress and establish a new routine (see page 43).
- **Keeping a journal** allows a person to express and explore feelings privately. A journal can't talk back, and can be an outlet and a comfort.
- **Books and movies** enable temporary escape from the ALS reality. When page turning becomes difficult, there are mechanical page turners, books on tape and books online as alternatives.
- **Soothing music** can have a calming effect when someone needs help getting to sleep or overcoming anxiety. Other music is great for stimulation and escaping depression.
- **Nature** never fails to provide something interesting and calming to watch, smell or hear. Focus on a hummingbird's flight, the breeze through a tree's leaves, the changing light during a sunset to find a moment's escape from problems.
- **Meditation** is a good coping skill for those with ALS and their caregivers. It can bring peace and relaxation

in the moment, and over time can help one develop a more open and accepting view of life. The practice of meditation doesn't require any specific spiritual belief, and can increase physical energy by dispelling stress. Practiced as little as 10 minutes twice a day, it can improve a person's overall well-being.

For more information:

- “Adjust, Adapt and Cope,” MDA/ALS Newsmagazine, June 2007
- “ALS: Anyone’s Life Story,” MDA/ALS Newsmagazine, May 2007
- “All in the Attitude: Determining Your Quality of Life,” MDA/ALS Newsmagazine, August 2006
- “ALS Caregiver Suggestions: My Perspective,” MDA/ALS Newsmagazine, July 2005
- “Survivors Tell Health Care Professionals: Give Us Hope,” MDA/ALS Newsmagazine, July 2004
- “The Survivors: What Keeps Them Going,” MDA/ALS Newsmagazine, December 2003
- “Readers Respond — Is ALS a Disability or a Fatal Disease?,” MDA/ALS Newsmagazine, February 2003
- “ALS — A Disability or a Fatal Disease?,” MDA/ALS Newsmagazine, January 2003
- You’re Stronger Than You Think, Tapping Into the Secrets of Emotionally Resilient People*, by Peter Ubel, M.D., McGraw-Hill, 2010 (available as an e-book only)
- Journey Beyond Diagnosis*, by Greg Pacini, Reedy Press, 2005
- The Art of Getting Well: Maximizing Health When You Have a Chronic Illness*, by David Spero, R.N., Hunter House Publishers, 2002

Anger

Anger is a more-than-reasonable response to ALS. This disease is a cruel thief that no one can stop. No one deserves it.

But ongoing anger leads to damaging stress, prevents acceptance and can lead to abuse or unhealthy behavior.

Sitting on anger and trying to hold it down usually is ineffective. Some people think that if they open up and let the anger out it will take over forever, but that's unlikely to happen if approached in the right way. Go somewhere safe and

“I feel that writing down your feelings, thoughts and ideas on a daily basis is important for anyone, but most important for family caregivers. I remember driving home from the long-term care facility in which my grandfather was living and flashes of the times we would have together when I was a kid would pass quickly before my eyes. I couldn’t wait until I got home to write them down.”

Studies show that when people write about emotionally difficult events or feelings for just 20 minutes at a time over three or four days, their immune system functioning increases. ■

— “Today’s Caregiver,” Gary Barg, editor-in-chief, todayscaregiver.com

private — the shower, the woods — and feel the anger, let it out physically. Scream about your hatred of ALS and anger at it. Physical exertion such as running or hitting a punching bag also may help dispel some anger. One caregiver’s friends gave her a “damnit doll” made of cloth and stuffing that she could whack on a table whenever she couldn’t contain her anger.

If anger makes you yell or snap at people, apologize and explain that it’s the disease you’re angry at.

The person with ALS also gets angry at the disease; at the loss of physical ability, independence and control; at exhaustion, dependence and every specific loss. Show patience so your loved one can see you coping and continuing to regard him or her as the same person. People with ALS may strike out in anger if those around them aren’t patient enough to understand what they’re trying to communicate, or to take time to help with physical tasks that may take many hours. Being unable to “fix” things also stimulates anger.

For people used to making decisions or controlling others, or those who strongly value their independence, anger may be a primary response to ALS. The caregiver’s acceptance of this anger will help the person gradually become more cooperative with the caregiver, family and the medical team, and stop fighting those who are helping.

“ALS is hard for those of us who are used to being independent and in control of ourselves and how we live our lives! My family has, more often than I care to admit, taken the brunt of my frustration — especially if I am tired (which is most of the time). My fuse is amazingly short and patience is nonexistent when I am tired. **”**

In childhood, some males are taught that anger is the only acceptable emotion, so it may be the only one a man knows how to express or even recognize. Similarly, females may have been taught that they never should show anger, and in adulthood a woman may have trouble even realizing she's angry, and have to learn ways to acknowledge and express it.

Caregivers may feel angry for the same reasons as the person with ALS. In addition, they may feel angry at the loved one at times, no matter how devoted they are, because they feel abandoned or burdened. These feelings are normal, and they're part of a displaced rage at the disease; in time, they'll be incorporated into a wiser, more patient view of the situation.

Fatigue — a familiar state for caregivers — shortens patience and control. Try to make it clear that you hate ALS, not the person with it. If anger becomes overwhelming, look for counseling or care assistance before it escalates into abuse (see page 96).

Manage anger by gathering information about ALS and what can be done about it. This decreases helplessness and gives specific tasks to focus on instead of anger. Forgive yourself for having some crabby days; it's only human. Other anger management strategies include: practicing relaxation techniques such as meditation; talking with a trustworthy friend or counselor; or including enjoyable activities such as shopping, movies or sports in your schedule. There's nothing like focusing outside yourself to dampen the intensity of your feelings.

Anxiety

Anxiety is a feeling of apprehension and fear often characterized by physical symptoms such as palpitations, sweating or tension. Anxiety, which may arise at the time that denial is breaking down and the reality of ALS is setting in, also may manifest as restlessness, fear of suffocating, fears of specific situations, trembling, shortness of breath, dry mouth, dizziness, nausea, irritability, obsessive worry, loss of sleep and inability to concentrate. In an anxiety or panic attack, these sensations take over and persist for more than a few minutes.

In a person with ALS, panic or anxiety sometimes is related to breathing issues or position; (see pages 51 and 65 to address these issues).

For caregivers, stress can bring out anxiety symptoms as events seem overwhelming. A person may feel abandoned, endangered, very vulnerable. Deep breaths, calming and assuring words, and recognition that this is a passing sensation, may help get through the immediate crisis.

Continued anxiety may require periods of respite for the caregiver; and, for both caregiver and the person with ALS, counseling or anti-anxiety medications such as lorazepam (Ativan), alprazolam (Xanax) or diazepam (Valium). Some antidepressants also have anti-anxiety effects, while others, such as bupropion (Wellbutrin), can actually increase anxiety. Talk to the doctor to figure out which will work best.

Respite for a caregiver can be a weekend or few days away, or it can be as simple and brief as taking a short walk, playing some favorite music, gardening, taking a bubble bath, shopping, meditating, or getting a hug from a friend. Learn to recognize when you need a break; even 5 minutes here and there will help.

Attitude

“Keep a positive attitude” is a cliché, and, like most clichés, it begins in truth. Any time a caregiver or care receiver can focus on finding solutions, and believes a solution is at hand, emotions will be less extreme, and stress will be abated.

Yes, you can change your attitude. Thoughts affect emotions; that's the basis of cognitive therapy offered by psychologists. By redirecting or adjusting the things you say to yourself, you can alleviate some stress.

Obviously, it's necessary — and unpleasant — to face the reality of ALS, and to do realistic planning and problem-solving. But when stressful, negative thoughts become obsessive, and you frequently find yourself overcome with negativity, it's time to change your thoughts for your own mental health.

For people with ALS and/or their caregivers, negative thoughts may fit into three categories:

- Reminding yourself of the disease and that the loved one will never recover
- Thoughts about limitations and losses
- Fantasies of terrible things that await in the future

These thoughts can be altered or shut off. When you notice yourself caught up in negative thinking, consciously change the message you're giving yourself. You might say to yourself, "I'm going to stop thinking about this and instead I'm going to think of (or do) something else." Or you might challenge yourself to "look for the good in this," or to explore some solutions.

Remind yourself that no one knows for sure what the future holds, and that future events rarely turn out to be as bad as imagined. Consciously bring your thoughts back to the present.

Many people find relief, gratitude and renewed strength in counting their blessings at such times, not overlooking even the smallest one. Or you might simply start singing a favorite song to put your mind back in a positive place.

You aren't in denial — ALS is still there and you're still dealing with it. But you're strengthening your positive attitude and re-affirming the belief that "together we're going to have a life that matters, a life to enjoy, despite this disease."

Some physicians, backed by studies, believe that the positive attitude or "fighting spirit" of a person with a medical crisis helps diminish symptoms or increase survival. Faith

(see page 112) that there's a purpose for the disease, or that you'll have a good life despite it, can contribute to a more peaceful experience. Acceptance, or by contrast, determination to defeat the disease, all can be emotionally — and sometimes physically — strengthening.

Burden and rewards

Caregivers may say, "Oh he's no burden. I love him," and mean it. But caregiving *is* a burden, or rather a huge pile of burdens. The load can be very hard to carry at times, and every caregiver has days when they just can't find the strength. It's better to acknowledge these times than to pretend you can do it all or that it's no trouble.

Smart caregivers get all the help they need to manage the chores and responsibilities that caregiving brings. Throughout this book, there's information about equipment, medication, medical procedures, agencies, people and other resources that can help, especially in Chapter 8. Always remember that it's ALS putting the burden on you; your loved one isn't the burden.

The burden is a bit lighter for caregivers who can unload resentment, anger, guilt and other negative emotions. Exhaustion, fatigue and stress will occur at times. Don't minimize the emotional and physical demands of the caregiving role. Fatigue weakens the body's immune system and makes caregivers more susceptible to illness. Poor eating and sleeping habits, stressed joints from lifting, and the risk of developing serious illnesses such as a heart condition or high blood pressure have been documented in longtime caregivers.

There are benefits to long-term caregiving as well — greater intimacy, increased patience, knowledge that you're needed, and gratification that you're making someone you love happier, secure and comfortable. A recent study of caregivers of people with Alzheimer's disease showed health benefits associated with helping others, especially when the caregiver and care receiver share a close relationship. "Caregiving is often a joyful and beneficial experience for the one who gives," the researcher found, especially "when provided with proper respite and support."

Caregiver emotions and stress

In some ways, ALS is harder on the primary family caregiver than on the person with the disease. One study showed that, over several months, family caregivers of people with ALS reported a significant increase in depression and a sense of being burdened. In contrast, their loved ones with ALS didn't report a change in their quality of life or an increase in their level of depression. Lack of time for oneself was the main source of caregiver dissatisfaction.

The big umbrella over all caregiver emotions is *stress*. Stress occurs when there's a perceived discrepancy between the demands of the situation and the ability to meet those demands — *I just can't handle all this!* It's made worse by fatigue, worry, ever-increasing demands, illness — and it's an inevitable part of caregiving.

Stress in small doses actually increases energy and the drive to get things done. But it can become overwhelming to the point of illness or inability to give care.

Signs of stress include:

- inability to concentrate
- unexplained crying
- short temper
- physical symptoms like pain or throwing up
- nightmares
- sleeplessness
- irrational fears
- nervous habits
- constant worry
- mood changes
- sweaty palms

(Stress has some of the same symptoms as anxiety (see page 100), but in anxiety there's an overwhelming sense of fear or dread.)

A family may have unique circumstances that add even more stress to the daily tasks of ALS caregiving — other conflicts, illness or disability in the family; financial problems; abuse; family disagreements about care; family members far away or kids preparing to leave for college.

Even happy changes like weddings, graduations or births can feel bittersweet, or can add to the burden of chores.

If a caregiver is holding a job, raising children, concerned about aging parents and taking care of someone with ALS, the day simply isn't long enough. Sleep may become a luxury, making caregiving harder and stress greater. (See "Sleep deprivation," page 57.) Despite exhaustion, sometimes the mind can't slow down even in bed and the caregiver lies awake. Stress has reached the boiling point when the caregiver can't sleep for several nights in a row; gets too sick to perform the duties; takes out frustrations on others, including the person with ALS; or just "falls apart."

But, as one caregiver pointed out, the mental and emotional anguish felt by those with ALS and their caregivers is the one ALS symptom that can be reversed, or at least controlled to a large degree. Another caregiver says the keys to getting the job done are *preparation, flexibility and a sense of humor*. When you've lost those capacities, you've got to R.E.C.H.A.R.G.E.

R: Rest.

E: Eat right.

C: Communicate your needs to others.

H: Hydrate. Drink lots of water.

A: Accept help.

R: Respite.

G: Get enough sleep.

E: Exercise.

It's OK to let the loved one know that you're stressed; try to do so in a way that doesn't provoke guilt for being the cause of the stress. Make it clear that ALS, not the person you love, is the source of exhaustion and pain. Your loved one may be able to console and support you — with thanks, with ideas — and that can go a long way toward lightening your load.

One caregiver comforts herself with the thought that if she had ALS, she'd want her husband to give her as much care as she's giving him. She can do no less. Other ways to manage stress include:

- Find a support system.

- Schedule some time away from the person you're caring for.
- Practice relaxation techniques.
- Get medical help for sleeplessness, anxiety, depression or other painful emotions.
- Acknowledge the enormity of the caregiving responsibility and take credit for doing it well.
- Stay away from other people who increase stress, if possible.

Here are some specific techniques to help with sleep and relaxation of the mind:

- Fill your bathtub ankle deep with cool water and march around in it for a few minutes.
- Visualize something peaceful like dancing butterflies.
- Try herbal teas or capsules of St. John's wort.
- Put thoughts into a journal, or start writing a poem to refocus negative thoughts.
- Wiggle your toes.
- Listen to a tape or CD with sounds of nature like water flowing, rain falling and ocean waves rolling onto shore.
- Meditate.

“My mind is like a puddle right now. I pray and pray some more for strength and guidance. At night it's so hard when I come home, not being able to converse with anyone. I do everything I possibly can to keep it together. My wife is my first priority, but Lord knows how hard this is getting.**”**

“[My husband] surprised us all with his ability to cope with and adapt to monthly, weekly and sometimes even daily changes, and maintain a positive attitude. In the meantime, I dealt with everything in my own way — with chocolate and humor — as I discovered time wasn't my own but belonged to caring for him in every way.**”**

Here's another simple relaxation procedure that takes only a few minutes:

1. Find a comfortable, quiet place. Sit with head, arms, legs, neck and back supported, shoes off and clothes loosened. Close your eyes.
2. Take a deep breath and hold it. Be aware of the muscle tension in your chest. Slowly exhale and notice the chest muscles relaxing. Repeat.
3. Perform step 2 focusing on different parts of the body, starting with the toes and moving upward to the top of the head.
4. Slow your breathing even more. Let your mind wander to a peaceful image.
5. Remain in this relaxed state for several minutes.
6. Return to the present by counting slowly from 1 to 5.

If there isn't time to go through the whole exercise, the technique can help at odd moments during the day. Close your eyes and take a deep breath. Tense and relax the muscles in one part of the body.

For more information:

[“Caregivers May Be More Discouraged Than People With ALS,” MDA/ALS Newsmagazine, May 2007](#)

[“Writing About Events May Lower Caregiver Stress,” MDA/ALS Newsmagazine, February 2004](#)

[Health AtoZ](#)

[healthatoz.com](#)

Search for “caregiver” for several articles on maintaining health.

[Meditation for Beginners: Six Guided Meditations for Insight, Inner Clarity, and Cultivating a Compassionate Heart, by Jack Kornfield, Sounds True, 2004](#)

[Caregiver's Reprieve: A Guide to Emotional Survival When You're Caring for Someone You Love, by Avrene L. Brandt, Ph.D., Impact Publishers, 1997](#)

[The Miracle of Mindfulness: A Manual on Meditation, A Zen Master's Method of Meditation, Concentration, and Relaxation, by Thich Nhat Hahn, Beacon Press, 1987](#)

Children

The children close to a person with ALS emotionally respond to the diagnosis in their own ways, depending on their age and other factors in their lives. Each child's emotional process of reaching acceptance should be respected, and their questions answered in age-appropriate language.

Rather than simply telling children everything you know about ALS, neutrally bring up the subject and let them ask whatever is on their minds. You'll know how much information children are ready to hear by honestly answering their questions and then stopping. Sometimes they'll ask more questions; sometimes their curiosity is satisfied and they'll drop the subject. Take a moment to figure out what the child is asking. *Why can't Mommy come to my soccer game?* may be a question about ALS, or it may be a question about finding ways to spend time with Mom.

Children know when something's going on in the household, and they're disturbed when parents seem to be keeping secrets. Parents may want to wait until their own emotions are under control before discussing ALS, but it should be brought up when ALS changes become noticeable. Questions such as *"Why does Grandma fall all the time? ... Why can't Daddy ride bikes with me?"* mean it's time to open up the subject for discussion.

Young children

Explain the disease and the situation in simple terms.

Daddy has a sickness that makes some of his muscles weak. We may have to help him sometimes when his legs and arms aren't strong.

If they're interested, you can tell them the name of the disease and a simple explanation of how it works. A telephone analogy might help: *The brain calls up the muscles to tell them what to do; the message goes through nerves, like telephone wires. Sometimes if the wires are broken, the muscles never get the message, so the muscles don't work.*

Follow the child's lead and don't explain more than they're asking to hear. Take your cues from the child as to how much he or she can digest at a time.



Speak matter of factly, and try to convey the overriding message: *We are all going to get through this together.*

Answer all questions at the child's level. In children under 10, the first reaction to hearing that a parent or grandparent has a disease is often fear. *Can I get it? Is he going to die? Will someone still take care of me?*

If the kids start to cry or show their fears, address those feelings honestly. *I know this makes you sad. It makes me sad too. But Dad is still here with us now, and he still loves us just the same. It's OK to cry when you're scared or sad. Mom and Dad cry about ALS sometimes, too.*

Give reassurance that no matter what happens, they are loved and always will be cared for. The outside things may change (such as the loss of physical abilities) but the inside feelings won't change.

Show that you're strong enough to talk about ALS even if it makes you cry sometimes. This prevents kids from thinking they shouldn't ever bring it up for fear of upsetting you, and lets them know it's OK for them to cry too.

If a child is caught up in tears or shies away from the loved one, it might be due to fear or guilt. *No, honey, you can't catch ALS from Grandpa. And he didn't get ALS because of anything you or anybody else did. It's not because you left*

your toys on the floor at his house. It's a disease and we don't know why it happened but sometimes it does.

Some kids may shrug off the news or seem uninterested. Let them process the information at their own speed. Although you can't force quiet children (of any age) to talk, you can let them know they're welcome to ask questions

“ My 11-year-old daughter is the one that helps me in the bath sometimes. She would not want it any other way. She wants me to know she is grown up enough to help me. My children really help so much that I don't know what I would do without them. We face new obstacles every day and we face them together. ”

“ In the back of my mind is the knowledge that my children are watching and learning from me every moment of every day.

I try to let that knowledge guide my behavior on the days when I want to hide under the covers and never come out. Is that how I want them to remember me dealing with adversity? Or how I want them to face adversity in their lives?

No, I want to teach them to dig deep and find the inner strength to live each moment to the fullest. ”

“ When I keep people from bumping into you, I feel like your knight in shining armor! ”

— Said by a child to a parent with ALS

“ At the age of 5 my daughter knew about ALS. She would fear going to school, as she thought I wouldn't be home. I kept promising her I would always be there.

I was there to see her graduate from high school. Now she's married and, went and all, I got to walk her down the aisle and dance with her at her reception. ”

or share feelings whenever they want. Take advantage of private times, such as when driving in the car, to offer them the opportunity to talk if they want.

Explain each new piece of equipment or change in the household as a way to help. Make it seem interesting and really wonderful (which it is!) that Dad has this wheelchair so he can go places. As each physical change occurs, remind them: *There are many ways we can help Dad, and lots of good equipment to help him eat and breathe and talk.*

Keep family life as normal as possible: Go to church, get the kids to school on time, visit relatives, go to restaurants, continue to discipline and express expectations of the children. Observe important milestones in the kids' lives — birthdays, school plays, soccer games — just as before, so they'll know they're not any less important.

Children pick up on their parents' tone of voice, unspoken feelings and actions as much as their words. Let them see the loved one participating in family life; the caregiver helping cheerfully; problem solving; and lots of hope and confidence. Suggest ways they can show their love — draw a picture, visit, make a craft, write a card/note, pick flowers, watch a movie together, etc. For a parent, helping around the house means a lot.

Children whose routines are disrupted may display developmental regression: need for a security toy or blanket; bed wetting; finger/thumb sucking; need for more frequent cuddling, etc. The response should be reassuring: *I see you're carrying Blankie again, and that's OK while you need it. When you don't need it, we'll put it away again.* Teasing or shaming the child for using familiar comfort objects only increases anxiety.

(Also see “Parenting,” page 124, and Appendix C, “Talking to Your Kids About ALS.”)

Older children and teens

Older kids, 10-18, will feel some of the same fears as the younger ones. However, they'll have a better understanding of the disease and awareness of its seriousness.

They may resent not being the center of attention, or conversely, may want to help as much as possible. More than a million American children provide some care for a chronically ill or disabled family member. Older kids can

My Yearlong Community Project Report

by Victoria Vanderpool

My yearlong project was to help my grandfather, General Tom Mikolajcik. He used to be a pilot in the Air Force. He helped design and later flew the C-17, a plane that carries cargo. But now he has a disease called ALS (amyotrophic lateral sclerosis, or Lou Gehrig's disease). It makes your whole body paralyzed. First his arms became paralyzed, now his legs are. Someday he will not be able to move anything but his eyes.

I help him by doing things that we do everyday; I comb his hair, feed him meals, give him water, help him go up and down the elevators, help him go through the newspaper when he reads it. I also do a lot of things that other people may do with their grandfathers like reading books together. I help him by turning the pages.

When I help it is a treat. I mean someday he will not be here and I will wish that I spent more time with him.

How does he move around? Well, he has a mechanical chair that he can drive with his fingers. He also has three elevators to get up and downstairs at his house. How

help with simple things like tying shoes or reading to the person with ALS. They also can help with more complex chores like cleaning feeding tubes or suctioning trachs.

Kids even may come up with some adaptations to make it easier to transfer Dad into the car or feed himself. Thank the child for being so thoughtful; use the suggestion if it isn't dangerous, and the child will feel the contribution is important to the family.

Take older kids to a clinic visit so they can ask questions of the doctor, nurse or social worker. That will acknowledge that their experience with ALS is important.

Sometimes a child expresses anger at the person with ALS, or says something like *I wish she could walk*. Just acknowledge that sentiment. *Yes, it really stinks. But that's the way it is in our family and we're making the best of it.*

does he bathe? Well, everyone has to bathe and he has a mechanical lift so he can get into the bathtub.

I like helping him. It makes me feel responsible and confident. I help my family community by helping him because usually my grandmother has to help him. Let's say it is lunchtime and I already ate. I would feed him so my grandmother can eat her lunch and she doesn't have to feed him.

My grandfather helps our community in a lot of ways. So if I help him, I help our whole community. If we just left him in his chair he would die because he cannot feed himself or even get up. My family will try to make his life as good as we can.

I feel proud to have him as a grandfather. Having ALS may be hard for him but not him alone because many other people suffer from it, even [S.C.] Governor [Mark] Sanford's father died from ALS.

I think that what I chose to do this year was a good project. This isn't just a yearlong project ... it is a life-long project. I love him!!! And that's what my yearlong project is.

Victoria was a 10-year-old fourth-grader when she wrote this school report. She also did a PowerPoint presentation using photos of her grandfather.

Not infrequently, older children feel embarrassed about the way the person has changed physically, or when people stare at the power wheelchair, vent mask or communication device. Suggest that really close friends will understand if you tell them about ALS and explain that, underneath, the person is just the same as he or she used to be.

Life and death questions don't have to be addressed at the beginning, but at some point a child — especially one who's done his own research — will want to know if the loved one is going to die.

A possible answer: *No one gets completely well from ALS, and sometimes it gets dangerous because it keeps people from breathing. We're going to do everything to help Mom, but someday her body may get so weak she can't keep going. Right now, Mom still wants to help you with your home-*



work and hear about your day. It makes her really happy when you spend time with her, and she loves knowing that you're having fun and doing things you like.

Encourage kids to talk to adults about their fears or worries — either their parents, an aunt or uncle, or someone else they trust. Ask at the ALS clinic about family counselors who can see family members individually and as a group.

Adult children

Most family caregivers are spouses, but sometimes an adult son or daughter becomes the primary caregiver of a parent with ALS. (At other times, a parent must care for an adult offspring who has the disease.)

The need to care for a parent can be very disruptive to a busy adult's life. Just when a person wants to give attention to children or career, or to enjoy the prime of life, they're pulled back into the old role of child, but with more complications. An adult child may have to relocate and set life aside for a while. That's a lot to ask, but no

one else can be sure a mother or father is cared for the way their own child can. (For those who can't relocate, see "Long-distance caregivers," page 161.)

Parents and children often reverse roles as the parent ages, and there'll be some role reversal in ALS even if the parent is only middle-aged. To the extent possible, savor the time with the parent, and live day by day. Find out how she'd like to spend her time and plan some fun things to do together while she's still able. The parent will appreciate the devotion, and children can be glad to return the care they received in childhood.

Caregiving situations can highlight unresolved conflicts between parent and child. Distrust, disrespect or resentment may be part of the caregiving experience. Knowing that this time is precious for both parties, that now is the time to forgive and forget, can help do away with old business. Try not to relive old conflicts, but lightly say, "I guess I was a handful sometimes. I'm sorry if I hurt you." That may be all that needs to be said.

Some parents will find it ludicrous that their child is telling them what to do, so press advice gently. Remind her that the doctors at MDA clinic said she'd be better off using a walker, or a feeding tube. She may resist at first but eventually come around. The adult child may have to work for a while to get the parent to open up financial records and discuss end-of-life plans; plant these seeds and say you need to know what he or she would want before a crisis strikes.

Humor helps in a lot of frustrating and difficult situations. Be openly affectionate with hugs, massages, foot rubs, hair brushing or shaving. These are soothing ways to communicate affection and tenderness.

In rare cases, a teenage child becomes primary caregiver for a chronically ill parent. Between high school or college classes, she'll rush home to provide care or see to the younger children. "I can't totally abandon my mom," one 18-year-old said. "She needs me." This huge responsibility can be incredibly stressful on some young caregivers, leading to antisocial behavior, illness, isolation and postponement of dreams. Other teens blossom with the responsibility; this is more often the case when parents make sure the teen still has time to be a teen as well as a caregiver.



If you're in that situation, see the social worker at your MDA clinic and ask where you can find help. There should be support through the social services system, which someone familiar with ALS can help negotiate.

For more information:

See "Parenting," page 124, and "Guardianship," page 145, and Appendix C, "Talking to Your Kids about ALS."

"Having Children After an ALS Diagnosis," MDA/ALS News-magazine, July-August 2010

"Role Reversal: When Children Care for a Parent with ALS," MDA/ALS News-magazine, November-December 2008

"When a Parent Has ALS: Tips for Helping Children Cope," MDA/ALS News-magazine, March 2008

"Kids and ALS," MDA/ALS News-magazine, April 2005

My Grampy Can't Walk, by Vanita Oelschlager, Cleveland Clinic Press, 2006

Bigger Than the Sky — the True Love Story of Emilie and Her Grandfather, by Helene E.D. Nichols, Xlibris, 2003

ALS — Lou Gehrig's Disease, by Mary Dodson Wade, Enslow Publishers, 2001

“To know that he is better now than he will ever be again, and to make wonderful memories together, to spend lots of time loving and living together, to make each day special, because each day with him is precious — those things are important. **”**

“Whenever I hug my father, it reminds me how safe I felt whenever he hugged me. I always knew that no matter what happens in this world, my dad will be there to protect me.

Now that I am here and have literally stepped into his shoes, I wonder if it will ever be the same again. He may never be able to just hug me and say, 'All will be fine, I am here so don't you worry about anything' or 'Do you need any money?' **”**

Are You Tired Again? I Understand: An Activities Workbook to Help Children Understand and Live with a Person Who Has a Chronic Illness or Disability, by Marilyn Weisberg Deutsch, Ph.D., Western Psychological Services, 1998

Why Do Some People Use Wheelchairs?, Dorling Kindersley Publishers, 1997

ALS Society of Canada.

als.ca/als411

Child friendly resources, including booklets for children, teens and parents such as "When Someone Special Has ALS."

"Telling Kids about ALS/MND"

alsindependence.com

Dementia

Although older literature suggests ALS doesn't affect cognitive function, a growing body of evidence indicates that thinking and behavior change in about half of people with ALS at some point during the course of their illness. Usually (although not always) these changes are subtle. (See "Cognitive changes" in Chapter 2.)

Uncharacteristic thinking or behavior can indicate a condition called **frontotemporal dementia (FTD)**, which occurs because of ALS-associated changes in the frontal and temporal lobes of the brain. Sometimes the person with ALS has such extreme moods and behavior that he or she is abusive or unmanageable or may seem to be a different person. (See “Abuse” and “Anger” in this chapter, and Appendix B, “When the Thinking Parts of the Brain Go Awry in ALS.”)

When dementia is suspected, try not to take outbursts personally. The person with ALS has no more control over this behavior than over breathing or swallowing problems. One caregiver whose loved one frequently accused her of incompetence handled outbursts this way: “I agree with him that I certainly have my shortcomings, but I reassure him that I do love him and I am trying. I try to be an instrument to show him love without condition and just for who he is, despite his behavior.” Trained home health aides may be able to help manage the person’s behavior.

It may be impossible to maintain the person at home if the behavior is damaging to the caregiver’s or family’s well-being and safety, or results in destructiveness. See Chapter 8 for information about nursing homes.

For more information:

“Frontotemporal Disorders: Information for Patients, Families and Caregivers” (2010 booklet), National Institute on Aging (National Institutes of Health), www.nia.nih.gov, (800) 438-4380.

Denial and hope

Denial usually is the first stage of grief, and it serves a purpose. Early in a crisis, thinking “This can’t be true” helps ease the mind from shock, while the reality slowly sinks in. A degree of denial can be a way of holding on, by looking for the hopeful message in any news — *maybe it won’t be as bad as they say; maybe there will be something to help or fix this problem.*

The determination to find a cure, or to be the one who survives against the odds, could be seen as denial. But this kind of determination also fuels and arms people for the daily fight. If a focus on finding a cure causes caregivers to ignore daily needs, then it’s gone too far. Denial is the opposite of acceptance, the stage of grief

“*Mom is stubborn! She will only allow certain people to help her and even then she only allows us to do certain things. This makes it much more difficult to handle because I see her and her caregivers struggling more than necessary. I have to respect her need to hang on a little longer, even if that means more work for us.*”

“*My sister has ALS and is progressing pretty fast. My parents won’t accept that this is what she has, even though she has been diagnosed by two neurologists, and they won’t go to any local support group meetings.*

My parents say that when [my sister] is asleep she is able to move her legs and roll over in her sleep so she can’t be paralyzed so it isn’t ALS, she just doesn’t want to try. I think my parents are full of it.”

or crisis management in which people can function and deal with problems.

As with most psychological states, danger occurs when denial shuts out the possibility of emotional acceptance. If a patient or caregiver refuses to admit that the person has ALS, they may neglect to take the steps to ensure proper care. Denial can keep a person from using a wheelchair or ventilator until great, irreversible damage has been done. It may keep the person from planning for guardianship of children, making a will or advanced medical directive, or discussing assisted ventilation options. A person in denial can’t make a life with the disease, and the denial doesn’t affect the disease course.

When family members can’t accept the diagnosis or the need for assistive equipment and life changes, conflict is set in place. Those in denial may accuse the person with ALS of not trying, may seek out unqualified medical opinions that agree with their interpretation, or may refuse to accept support or information. This kind of denial usually occurs because accepting the diagnosis is too painful; eventually they may come to accept it. Many experts — social workers, hospice teams, other

families with ALS — have experience dealing with those in denial. They can help guide family members to the truth while maintaining hope.

Denial is a normal, initial part of the grieving process, and it's healthy to move past it in stages, as each aspect of the disease is accepted. It's also reasonable always to hold onto hope, although the things you hope for will change over time.

Caregivers often deny how hard their role is. Wanting to do the best they can for someone they love, they believe they can hold onto their jobs, give appropriate attention to children and family, and provide care to the loved one with ALS, no matter how much is required. This is a formula for burnout. Experienced caregivers always emphasize: *Get the help you need*. Getting help doesn't mean your contribution is any less important or that people will think you're lazy; it shows you care enough to plan for the long haul.

Moving out of denial doesn't mean accepting the worst prognosis. ALS isn't completely predictable, and in general people are surviving longer with it. Keep up hope; it's a necessary part of survival with a good quality of life. Knowing that research is progressing, that some people reach a plateau with ALS, that some survive for decades, that eventually you will reach a new balance in your life, can provide strong emotional support on the ALS journey.

For more information

"Life Satisfaction Similar With and Without Disability," MDA/ALS Newsmagazine, July-August 2007

"Building Hope One Brick at a Time," MDA/ALS Newsmagazine, October 2006

"Staying Alive — Does Personality or Belief Make a Difference?" MDA/ALS Newsmagazine, November 2004

Depression

Depression is an organic condition involving brain chemistry. It affects thoughts, moods, feelings, behavior and even physical health. Sometimes it occurs for no apparent reasons; other cases, called *situational depression*, arise when bleak circumstances occur in someone's life. Depression can be a coping style, or a stage of grief in which people get stuck. About 10 percent of Americans have *clinical depression*, which needs medical attention. Among longtime caregivers, the incidence of clinical depression is at least twice that high.

Weakness of facial muscles may make a person with ALS appear expressionless or depressed, no matter what real feelings are going on. Other forms of communication are needed to find out if the person is really depressed or simply can't smile or show true feelings.

Oddly, some studies have shown that people with advanced ALS are no more likely to have depression than people who don't have the disease. Caregivers are more likely to be hit by depression than those they're caring for. Nonetheless, many people with ALS do require antidepressants. Watch for these signs of depression in the person with ALS *and* the caregiver.

Signs of depression include:

- persistent sad, anxious or empty mood
- diminished ability to enjoy once pleasurable activities or people
- restlessness or irritability
- quick agitation or anger
- changes in sleep patterns
- changes in appetite or eating patterns
- unexplained crying spells
- loss of sex drive
- decreased energy level and constant fatigue
- slowed movements
- difficulty concentrating, remembering or making decisions
- feelings of sadness, hopelessness, helplessness, worthlessness

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- ongoing physical symptoms that don't respond to treatment, such as headaches or digestive disorders
- recurrent thoughts of death or that life isn't worth living

In such a state of mind, the demands of caregiving easily will overwhelm anyone. A caregiver may be unable to summon the will to act. If any combination of these feelings lingers for more than a few weeks, get help. To fight off mild depression or "down" moods, try exercise,

“I didn't realize how much water the human body really does have in it, but I do now, as I find myself crying and crying all the time. **”**

“The antidepressant doesn't totally take the sadness away but it helps me cope with all that is on my plate now and to be able to keep going. Getting family and friends on board and keeping them updated helped a lot. **”**

“Sometimes I would awaken to hear him softly sobbing. At those times I usually didn't try to comfort him or stop him as I felt it might relieve some of the devastation he was feeling and trying not to show in the daytime. **”**

— **Caretaking** by June Suda,
Divine Light Publishing, 1992

“When [my husband] was diagnosed and life got rough, I asked my doctor for something and he refused. It took a few years but he finally apologized to me and put me on 10 mg of Celexa, which helped me rest at night and cope better during the day.

My doctor said now it has become his common practice to offer a caregiver medication when a patient is put on one. He now realizes, thanks to [my husband's] case, how tough an illness can be on those closest to the patient. **”**

laughter, friends, escape from routine, an enjoyable activity, counseling. If those don't work, ask the doctor about antidepressants.

The good news is there are many excellent antidepressants in the physician's arsenal. They may be the only way to blast oneself out of inertia and return some energy and zest for living. Antidepressants may take 10 days to six weeks to become fully effective.

Doctors can go over the various types of antidepressants and help decide what works best. Some have side effects; sometimes people need to try one, then switch to another, until the best choice is found. Some that people with ALS and their caregivers have found to be helpful are Lexapro, Zoloft, Prozac, Wellbutrin, Celexa and Lyrica.

Research indicates that a combination of medication and counseling are more effective against depression than either alone. Counseling can help a person talk over or resolve issues about the meaning of one's life, relationships or past actions. Many people take a round of antidepressants and feel better, then stop taking the drugs. In several months, they spiral down again. This isn't a failure — this is the nature of depression. If you need to take them a second time or for several years, do so.

Do whatever it takes to return some joy and energy — caregivers and people with ALS need all of those commodities they can find. The effect of depression is loss of any enjoyment of life, and thus loss of the will to live. It isn't something to take lightly.

For more information

[“Depression Doesn't Dominate in ALS,” MDA/ALS Newsmagazine, September 2005](#)

[“Reducing Caregiver Stress May Help Loved One's Depression,” MDA/ALS Newsmagazine, February 2003](#)

[“Tips for Caregivers: Handling a Loved One's Depression,” MDA/ALS Newsmagazine, September 2002](#)

[“Living Longer Depression Free: A Family Guide to Recognizing, Treating and Preventing Depression in Later Life,” by Charles Reynolds, Johns Hopkins University Press, 2003](#)

[“Chronically Happy: Joyful Living in Spite of Chronic Illness,” by Lori Hartwell, Poetic Media Press, 2002](#)

“So they changed ‘as fate would have it’ to ‘as faith would have it’ and thereby began reconstructing their life. **”**

— **On Any Given Day**, by Joe Martin and Ross Yockey, John F. Blair Publisher 2000

“My relationship with God is fundamental, but I don’t like the idea that you postpone all your payoff until you die. We start making our heaven or our hell right here.

I don’t pray for cures, I pray for healing and purity of heart, and if a cure comes, that’ll be wonderful. **”**

Understanding and Overcoming Depression: A Common Sense Approach, by Tony Bates, Crossing Press, 2001

Depression and Physical Illness, by Andrew Steptoe, Wiley, 1997

Faith

Faith is a great help in living with ALS. Faith can come from a traditional religious belief or from a belief in a well-meaning universe, the cycle of life, 12-step groups or any other school of thought that helps one make sense of life. Faith, like other states of mind, can’t be imposed on a person, but many people going through a crisis like ALS find themselves returning to prayer or other religious practices they’ve abandoned, or exploring their thoughts to recover their faith.

“Having faith” doesn’t mean believing in miraculous cures. It’s an internal experience that provides comfort, strength and meaning, and a place to turn when things are hard.

For more information:

Including People With Disabilities in Faith Communities, by Erik W. Carter, Brookes, 2007

National Volunteer Caregiving Network
(877) 575-4932
nvcnetwork.org

Family

Tolstoy wrote that every unhappy family is unhappy in its own way. Just imagine what happens when you add something like ALS to the complicated world of a family. When ALS is diagnosed, it’s in the context of the person’s immediate life and family, including extended family and other relationships. Everyone in that family will have a personal experience of grief and adjustment, going through many of the emotional difficulties described in this chapter. A crisis like ALS usually brings out people’s true colors. If it’s a normally cooperative family, they’ll work together to face ALS; if there are conflicts and competitions, those will dominate relationships and attitudes.

One person — usually the spouse, though sometimes a parent or an adult child — becomes the primary caregiver of the person with ALS. Expecting every member of the family to agree with and support the caregiver’s decisions and methods isn’t realistic. It’s said that families rally together in a crisis, and sometimes they do. It’s probably more truthful to say that family members relate to each other on the ALS journey in the same ways they’ve related before.

You may find support and love in unexpected corners, and some relatives will be wonderfully helpful. Extended family members also may offer constant criticism, unwanted advice, denial or “miracle” cures; they may refuse to learn anything about ALS or simply be unable to understand the information; they may resent the new friends and support



“His family wanted the vent turned off — when he was still writing and signing and laughing. They didn't think there was any reason for him to live any longer. He told them he did not want it turned off. They were the family from hell. **”**

“My folks visited frequently; my dad would sit with [my husband] while I ran errands, and every time he was hospitalized they would come and be with us. 29 times! That's family, that's devotion. Even the out-of-state relatives would call on a weekly basis, send small gifts and show concern. **”**

systems the person with ALS and caregiver have found. Among the worst responses, family members may refuse to assist with caregiving, and even sabotage the system the primary caregiver has worked out. They'll never visit, or even break off communication. An exhausted caregiver can feel powerful resentment and anger at this hurtful response. The only constructive thing to do is remind them that the person with ALS especially needs their love right now, then let go of any expectation of cooperation.

Be grateful for those family members who are willing to help, and forget the uncooperative ones. Sometimes an in-law or sibling is determined to oppose the caregiver, for some longstanding reason or because they simply can't face the reality of ALS. Forgive them and get on with your life. It's not uncommon for caregivers to get more support from friends than from family members.

Also see “Children,” page 104; “Friends,” page 114; and “Marriage,” page 120. See Chapter 8 for ways to encourage family members to help.

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Fear

ALS raises many kinds of fear — fear of losing control, of pain, of death. Fear that other loved ones will have the disease. Fear that the caregiver won't have the strength to cope, or won't know what to do.

People with ALS may fear that they aren't the same person as before the disease, and that others won't respect, need or love them if they change. Caregivers can reassure a loved one that they hate what the disease is doing but that their love always will remain, and that they'll always be there when they're needed. Caregivers may have to repeat this message many times in many different words, but gradually it will help calm fears.

Some caregivers constantly are afraid for the loved one's safety, concerned that a respiratory crisis, panic attack or choking fit may occur when their backs are turned. Reasonable precautions (see “Safety,” page 55) and good assistive equipment should minimize the possibility of a sudden crisis. If the loved one has a way to communicate when needing help — a bell or buzzer — the caregiver can be reached quickly.

“My dad's brother and sister-in-law were here for the weekend. We stuck around because they were scared and uncomfortable to be alone with my parents. It dawned on me that what has become routine and loving for us simply scares people that don't see him for weeks, even days at times. **”**

“One of my brothers came in for three hours yesterday — long enough to tell my Mom all she is doing ‘wrong,’ to let her know that she should be handling Dad and his ALS without help because that is the only way she will be able to live with herself after he is gone, that Daddy should drop the dignity issue because he is going to lose his dignity anyway, that she shouldn't need help in the house (she is 76 years old and hiring some sitters). He left her crying. **”**

“I lost my Dad to ALS after almost a year battle with it. Fear — I lived with it every day, almost let it consume me. Please try to let go of the fear. The fear will only beat you down. **”**

“When I see someone who is living the future that my hubby is going to have, it is a hard hit in the gut. I thought I was ready to handle everything that was going to come my way, but I'm scared for him and our future together. **”**

Fear can destroy a person by leading to worry, sleeplessness, panic or stress. The best antidote is information and a practical approach of dealing with each step as it comes.

Just recognize fear as it arises, and envision yourself picking it up and throwing it in a trash can, or walking around and past it. The less attention you give it, the less power it will have. Keep your goals in mind — solve the problem of the moment, enjoy something today even if it's only a look out the window at a blue sky. Turn to others in a support group or online group for reassurance. When the mind and hands are fully occupied, there's less room for fear.

Friends

Friends' responses to ALS will be varied. Many friends will truly care about your loved one's well-being but perhaps not know how they can help or relate to the person.

Some families are disheartened to find that friends disappear as ALS advances. There are several reasons why friends and family members sometimes seem to shun a family going through ALS:

- Most people aren't familiar with ALS, and don't know what to expect. They may feel too sad or upset to face a friend in a changed condition. They may fear the friend has become different, or that they'll lose their composure, or that they'll tire out the person with ALS. Many worry they won't know what to say.
- Friends from work may have nothing else in common when the person with ALS leaves the job.
- People who know a person as active and energetic may prefer to remember him or her that way.

- The person with ALS may be self-conscious about looking different to friends, or about a speech difficulty. He or she may discourage visits.
- Sometimes people want to help but are afraid they'll be asked to lift the person or assist in the bathroom.

These steps can help to allay fears and keep some friends involved:

- After the diagnosis is made, notify key friends and family members. Send them information about ALS or refer them to MDA's ALS website, als.mda.org, for information about the disease.
- Send out an email every month or so to report on the loved one's condition or activities. Use this as a place to list tasks for volunteers. MDA's *myMuscleTeam* care coordination website (mda.org/mymuscleteam) offers an efficient and effective way to harness the strength of your personal "Muscle Team" of family and friends.
- Remind people that their friend is still the same person — not someone who's sickly or unaware of surroundings. He or she may look, move and communicate differently, but has the same interests and cares about the same people as before.
- Suggest that, during a visit, friends talk about sports, politics, movies, children or anything else that interests them. They don't have to dwell on the disease. If speech is slurred or a communication device is being used, a friend who has the patience for a slow-moving conversation is a real gift.
- Invite friends to watch a ball game or DVD, to play board games or eat pizza. Activities present less of a conversational challenge.
- Friends can bring books, easy-to-swallow homemade dishes, or anything they think their friend might appreciate. Humorous books and CDs might be especially welcome. Emailing news and jokes can keep relationships going if visits aren't feasible.
- Schedule friends' visits so people don't come all at once and exhaust the person with ALS.
- Let those who want to help know there are many ordinary chores and errands they can perform. See Chapter 8 for more ideas.

“ Last year when we were on our cruise to Alaska, my best friend, her husband and my family did a ton of remodeling on the house. Widened doors, laid down hardwoods, started work on the accessible bathroom. All stuff I would have felt like I was imposing to ask for, but cannot tell you how appreciated it was. ”

“ At first when my husband was diagnosed and could barely get around let alone breathe, the people at his place of work were great, same with my fellow workers. For two years we had casseroles coming out of our ears, noses and eyeballs.

Now we have one, yes one, friend that still takes the time to come see him for an hour once a week. I value his visits for my dear husband as gold.

This is the one area where I blow my lid. If ever anyone of them should run into me and ask how my husband is doing, I will say, people with ALS are human beings that deserve respect, love and friendship. ”

“ I found that the more people that knew what was going on, the more support I received. ”

“ I could count on three or four friends who would ‘spell’ me, sitting with [my husband] so I could take a shower, run to the store or do housework. One friend would even come over, prepare dinner for us, then clean the kitchen — above and beyond the call of duty with teenagers in the house! ”

Caregivers can ask *their* friends to keep in touch in similar ways. Meeting for coffee or going to a movie gives the caregiver a break and keeps friendships going. The caregiver may have to take the initiative; friends may be unsure whether the caregiver has any time or what activities are appropriate.



Some friends will lose touch and social networks will dissolve. But friends are needed to fight off loneliness (see page 119), and new ones will be found. People who've been through similar ordeals or church groups may become more important. ALS support groups and other families met at MDA clinics will turn out to be important — they understand the ALS challenges and can offer specific support.

For more information:

Bedside Manners: A Practical Guide to Visiting the Ill, by Katie Maxwell, Baker Book House, 2005

Grief

ALS is a powerful loss for which grief is appropriate. Many families find the first six months after an ALS diagnosis are the most emotionally devastating time of the entire experience; in later stages there's more time to prepare for losses.

It takes time to accept the reality of ALS and get ready to play the hand you're dealt. Each person will experience this psychological journey on an individual timetable, and with various degrees of intensity. Sometimes the stages of grief — denial, anger, bargaining, depression, acceptance — occur in a different order, or recur several times.

Caregivers need to be patient as they and their loved ones and family members sort through these painful emotions.

Caregivers experience their own losses, beyond those affecting the person with ALS. They have less time available for other family members, and certainly less time for themselves. Caregiving is physically demanding and may lead to, or exacerbate, physical problems or illness. ALS often becomes the family's financial priority, overshadowing dreams of a new home, special vacations or a comfortable retirement, not to mention ordinary luxuries like movies or new clothes. Many caregivers must give up jobs or careers in which they've worked hard.

With unexpected changes in career, parenting, health and financial plans, the caregiver's life is turned upside down and redefined. Deep grief is natural; never think you aren't strong enough or shouldn't be feeling what you feel. Let the feelings come; cry and grieve the losses. Only after the pain is acknowledged can the caregiver fully focus on caregiving.

Even after one reaches some level of acceptance of the diagnosis, the emotional roller coaster continues. Every loss by the person with ALS or caregiver — walking, working, eating by mouth, breathing independently, changes in the relationship, conflicts with friends and family, financial distress, etc. — can bring its own powerful grief.

Knowing in advance that strong emotions will continue to pop up even when you think you're over them, can make the next bout a little easier and a little shorter than the first time. As you grow in experience and knowledge, you'll learn what coping strategies to use as needed.

For more information:

"In the Beginning," MDA/ALS Newsmagazine, February 2007

"10 Tips for New ALS Patients," MDA/ALS Newsmagazine, March 2004

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

“I felt like I cried for a year. The first year of diagnosis was really the biggest grieving time for me. Then I started to get busy preparing our house, trying to get the church on board, etc. Some thought I was rushing it but if you look back I was really right on schedule. Then I tried to make sure we were doing all the things we wanted to do. It certainly helps you to see what is REALLY important in life. **”**

“The ALS monster uses grief to throw rocks at us and sometimes we get blindsided and don't duck. **”**

Guilt

Feelings of guilt can be a response to any crisis. A poor relationship or unresolved conflicts can put guilt into the emotional mix.

Young children may feel they caused their parent or grandparent to get sick by "bad" behavior.

A person with ALS often feels guilty for imposing a hardship on the family or being a burden on a spouse. Be sensitive to these feelings; even jokingly calling the loved one a pest can trigger guilt. Let your actions show that you're in this together.

Caregivers may feel guilty for resenting what they have to do; for wanting a break from caregiving; even for wishing the loved one would go ahead and die so this will be over. Family and friends can experience survivor guilt for being able to eat a steak or run across the lawn when their loved one no longer can.

Guilt is the obnoxious cousin in the family of emotions. It serves no purpose whatsoever and is best ignored. Making mistakes is human; forgive yourself and get on with what you need to do. Apologize for serious wrongs, but it's more important to show love by current behavior.

Caregivers can reassure loved ones that they know they didn't get ALS on purpose and that they're caring and

How to Support Your Favorite Caregiver During the Holidays

Here are some “gift” ideas caregivers can mention to others, copy and post somewhere, or send to their church or club newsletter:

- Put up (or take down and put away) lights and decorations.
- Bring over a Christmas tree and set it up. Or take down the tree after the holiday and haul it away.
- Bake and wrap up treats the caregiver can give as small gifts to helpers and volunteers.
- Take the kids shopping for the caregiver’s present.
- Wrap and/or mail gifts.
- Take the car to get the oil changed or the dogs to be groomed.
- Offer to help type and copy a family letter to go out with holiday cards.
- Drop by to run the vacuum.
- Bring over a healthy casserole.
- Put up storm windows or shovel the sidewalk.
- Thoughtful caregiver gifts include: gift certificates for house cleaning, food delivery, yard work or pet grooming; a long-distance phone card; a meditation or exercise tape; a book of inspirational prayers; a handmade booklet of personal IOUs for such things as respite care, meals, chores, rides, dog walking.

Know that any time of year, emotional support and your time are the two most valuable gifts you can give a caregiver.

(Excerpted from the MDA/ALS Newsmagazine, December 2004)

helping out of love. This may seem obvious, but the person with ALS needs to hear the words spoken. Assure children that they don’t have the power to cause ALS, and are needed to continue to love and help the parent.

Holidays

Holidays, birthdays and other annual traditions present additional stress for people dealing with ALS. There are family traditions to uphold and reflections on the past year. Thoughts about time can bring up sadness and depression over the changes in the loved one’s condition and fear that this may be the last shared birthday, the last Christmas.

The year-end holidays can be especially stressful with all the shopping, special activities and visiting. Caregivers should look for ways to simplify the holidays and minimize stress, for the sake of the loved one’s physical health and the family’s emotional well-being.

Try these guidelines:

- **Prioritize.** Drop some holiday activities, and focus only

“ My wife and I have begun to pay strange men to see me naked. The worst part is, they don’t seem to enjoy it. I don’t enjoy it much either — and not for the reason my wife supposes, namely that with my ego I think they should be paying me. ”

“ One day my 3-year-old granddaughter asked her mother for a piece of candy. After three or four pieces of candy, she was told she couldn’t have any more.

Undaunted, my granddaughter continued to ask for candy, and continued to receive the same answer, ‘No more candy.’

Finally, in exasperation, my granddaughter announced, ‘I know, let’s pretend you are Grandma and you can’t talk!’ ”

“ Laughter is the only drug that works on ALS, and it’s free. What a bargain. ”

on the necessary ones. The entire house doesn't have to be decorated; a simple tree and wreath can be enough.

- **Simplify.** Travel can be eliminated if it's too hard. Let the relatives come to you and encourage them to stay in a hotel, or just include immediate family. Make simpler meals, or hold a potluck. Buy gift cards to favorite stores and activities, rather than trying to shop for elaborate gifts.
- **Avoid the mall.** Shopping online or via catalog saves time and energy and allows the person with ALS to be actively involved or to shop independently. And it keeps you and your loved one away from large crowds of sneezing, coughing, inconsiderate people.
- **No appointments.** Don't make any doctor, lawyer or other business appointments from mid-November until after New Year's, except in an emergency. Go when your schedule is less complicated.
- **Shun sweets.** Awww — this one doesn't seem fair! But exhaustion follows a sugar high. Eat healthier food, and spread the holiday goodies over time. If you're deluged with Christmas cookies, package some up and give them to others.
- **Stay in the present.** Focus on the fact that the family is together now. Do fun and special activities to make this holiday happy and memorable. Focus on the youngest in the family. When thoughts of the future arise, bring attention back to today.
- **Put resentments on hold.** Family conflict is a "holiday tradition" for many people. The holidays are a bad time to bring up problems. If conflict is fierce, keep relatives at a distance.
- **Be grateful.** Take a few moments to experience and express gratitude for the things you have: people, time together, shared love, the wisdom you've gained.
- **Ask for help.** Family, friends and neighbors often wonder how they can help or what gift would make your caregiving life easier. Tell them what you need: For example, ask someone to mail your cards or gifts, put up the tree, shovel the sidewalk, pay for a massage or a few hours of home health care.

“All my life I have seen myself as the provider for and protector of my family and friends. Now I need the tables turned, and I have had to learn how to say ‘Please help me’ instead of ‘What can I do for you?’ This may be the hardest adjustment of this whole ongoing process.”

[My husband] and I think one reason that holidays are difficult is because they're a poignant marker of progression. We're trying not to make any holiday such a big deal anymore (kind of hard to do with a 5-year-old) and instead to celebrate every day. I try to cherish the memories of the past but also live in the present.

The last Christmas [my husband] was with us, his caregiver and my youngest daughter decorated him to look like Rudolph! Red lipstick on the cheeks, red nose, funny antlers from the Dollar Store and a note that said 'I am lost. If you find me please return me to the North Pole.'

We took his picture and he thoroughly loved it! I spent that holiday with big round tree ornaments as earrings, just to add to the confusion.

Whatever puts a smile on your face ...

- **Flee the flu.** Lots of company and activities outside the house expose people to colds and viruses. Protect the person with ALS from these respiratory illnesses.

For more information:

“Tis the Season for New Holiday Traditions,” MDA/ALS Newsmagazine December 2008

“Holiday Stress Busters for Caregivers,” MDA/ALS Newsmagazine, December 2004

Humor

ALS isn't funny but life is. Great comedians know there's almost nothing that can't be laughed about. Laughter is a great stress reliever that's good for everyone.

Dents in the wall from a new wheelchair, strangers' weird comments about a person using a communication device, medical treatments and even falls can be seen humorously. Incorporating humor into your coping mechanisms helps a lot.

A man wrote a top 10 list of things having ALS had taught him, including:

- It isn't necessary to bite your nails.
- It isn't necessary to pick your nose.
- Some people with ALS have no sense of humor.
- There ain't no good in an evil-hearted woman.

Pets and kids can provide plenty to laugh at. Amusing TV shows, movies and books can be scheduled frequently to help lighten the mood. Even black humor has its place. There's nothing like laughter to dissolve a tense situation and bring people closer.



For more information:

"Tales from the Hydrant — Pets," MDA/ALS Newsmagazine, April 2006

"Laughing Along with ALS," MDA/ALS Newsmagazine, February 2006

"Fun with Frances: Tales from an Emergency Pet Shelter," MDA/ALS Newsmagazine, November 2004

"ALS: Tag Lines and Gag Lines," MDA/ALS Newsmagazine, June 2004

Loneliness

In addition to the demands on their time, physical energy and emotional resources, caregivers often feel very much alone. Only another ALS caregiver can fully understand all that's involved. It seems there's no time for old activities and friends. Caregiving for a spouse shifts the relationship to one of dependence rather than partnership.

Don't shut people out unnecessarily. There's a lot of help available, and many ways for isolated caregivers to connect and support each other. (See "Friends," page 114.) New friends made through MDA ALS support groups or online listservs may be crucial because they understand what you're going through and can give important advice and support.

Ask a good friend to give you a call at a scheduled time every week or so. That will keep you in touch with someone who cares. Also, take the opportunity to get out of the house when you can.

It may be difficult to listen to friends or empathize with their problems, when you're living daily with a disease as overwhelming as ALS. It takes an effort to offer compassion and resist comparing problems, but it's necessary for friendships to continue.

There are lonely nights and moments, but remember that others are going through the same thing. Some caregivers stay involved with MDA even after their loved ones have died, in order to help others going through the experience. Contact your local MDA office (800-572-1717) to be connected with others in your area who also are dealing with ALS.

Love

See "Children," page 104; "Friends," page 114; "Marriage," this page; and "Parenting," page 124.

Marriage

By far, the majority of ALS caregivers are spouses; more men get ALS than women, so wives more often are caregivers.

The vow to love "in sickness and in health" is put to the test when one partner has ALS. Many husbands and wives find that the experience strengthens their bond and becomes their life's most emotionally and spiritually meaningful experience, providing a level of closeness they never imagined. Love and devotion can fortify both partners, if they always remember that caregiving is an act of love. The greater one partner's needs, the greater the other partner's devotion.

Not that this always occurs easily or naturally. When the full reality of ALS sets in, and at other times over the years, the caregiving spouse may think of leaving. A caregiver can become frustrated and lonely from months and years of providing so many forms of care and not even having the gratification of spontaneous conversation. Sleepless nights, changed priorities, loss of equal partnership and other demands can cause resentment. Exhaustion can become overwhelming.

Some marriages don't make it. In those cases, it isn't ALS that ends the marriage, but other problems in the marriage that are made sharper by ALS. A successful marriage is difficult under the best of circumstances.

Changing roles

When ALS enters a marriage (or a long-term partnership), the nature of the relationship changes. Roles change; rather than two people dealing with ordinary life challenges together, you become two individuals facing a crisis individually and together.

At any time, spouses may be at different psychological stages with the disease, and each must respect the other's process. One may want to talk about the disease more than the other. Needs may clash; the roles and behaviors of other family members have an influence. Give and take, communicate

“Less than two months after I was diagnosed she stood before friends and family and said these words: ‘For richer or poorer, for better or worse, in sickness and in health.’ Never have these words meant so much.”

My wife married me knowing what lay ahead. It was an act of loyalty, love and yes, of courage, for which I will never be able to repay her.

It's hard to believe, but sometimes ALS stands for 'A Lucky Son of a Bitch.' ”

“Those times when it was one on one with each other when I would feed him were so comforting to me as his wife and caregiver. It was a warm and cozy time. Then as years passed and he became totally paralyzed and couldn't talk, our times alone were most precious to me. He loved for me to read to him. We read tons of books over the years, me sitting next to him, touching him while I read.”

“The pleasure I got, and the comfort knowing she was taken care of and she was in my arms and God was with us, took all the pain away. She said, 'I've never ever felt scared when you're around.' She made me feel like a king.”

“My husband is not the most gentle or affectionate man. He has rough, hard-working hands. I have worried about him being a caregiver as I weaken. I have been coaching him as to my needs, and bless his heart he is trying.”

So, much to my surprise, he had a big plan for my birthday in October. He was grinning as I came out and saw a 55-gallon aquarium set up in our living room. He said he knew how much I missed going snorkeling. He thought this way I could watch the fish all the time.”

and be patient, help partners be aware of each other's needs and get back on the same track.

As husband-and-wife shifts to patient-and-caregiver (or vice versa), it takes some effort to nurture the marital relationship. When one spouse depends on the other for virtually everything, how can two people maintain a partnership of equals?

- Give each other strength verbally and emotionally.
- See the disease as a shared experience. Don't compare who has it tougher. Sometimes one partner needs to lean on the other; at other times, the needs switch.
- Know the spouse with ALS would provide the same care if the roles were reversed.
- Don't infantilize the person with ALS. Respect his or her opinions and decisions even if the physical balance has changed. Avoid becoming a nurse or parent to your spouse.
- Do extra things to make the spouse comfortable, attractive and happy. Because of love, a spouse will provide the kind of care a paid caregiver can't give.
- Enjoy discovering more about each other as you spend more time together.
- Communicate in every way, especially to avoid misunderstandings.
- Continue to enjoy shared activities, including sex.
- Realize that today, the spouse is better than he or she ever will be again. Use the time to make wonderful memories, to share loving and intimate moments. Value each day together.
- Include others in the caregiving process so that some parts of the relationship can simply be between husband and wife.
- Keep laughing.
- Celebrate special times such as anniversaries.
- If resentment, depression or exhaustion seem to be taking over, look for some help. See "Caregiver emotions," page 102.

Intimacy and sex

Caregiving can provide for a special kind of intimacy. Intimacy isn't only sex — although ALS doesn't end sexuality.

Times of closeness and romance can come during special meals, dates or anniversary celebrations, or simply from talking, touching, laughing, hugging or sleeping together. A couple can enjoy the moonlight, favorite music or affectionate kissing. One woman found that feeding her husband was a comforting and cozy time for her. Reading to him, while holding hands, also became intimate and precious.

Touching, stroking, hugging, kissing and cuddling are needed by everyone; touching outside the tasks of physical caregiving helps keep the marital relationship alive.

Emotional intimacy includes:

- Acknowledgment of the other person's words and experiences
- Attention through listening, eye contact and nonverbal communication
- Affection through simple gestures like a kiss on the cheek or a back rub

Caregivers can verbalize some of their feelings to friends and others if communication with the spouse has become limited. Expressing and meeting some of these emotional needs on their own can help a caregiver focus more on the loved one.

Sexual relations often continue for people with ALS, as desire and function usually are maintained. Sex is an important form of intimacy and caring in marriage. The spouse with ALS may have an even greater need to be touched or caressed because their interactions have become so limited. Sexual release is also a great way to ease stress and facilitate sleep.

“Even though the method had to change somewhat, there was still desire. That never changed through all the physical changes. I had no problem separating the caregiving part and the wife part. I was his wife and lover first and caregiver second. **”**

Role Changes in Marriage

by Jo-Ann Goldstein

Every family is different, just as every progression and loss of function is different for each person with ALS. But we all face role changes, both as a person with ALS and as a caregiver.

Unfortunately, Sam's disease progression has been rapid and he now has no movement and very little speech. So, these thoughts are from my viewpoint. But we did discuss many of these feelings over the past two years and I know that I speak for him on some issues.

The biggest change we have had was for Sam to become the "taker" instead of the "giver" in the family. He had owned his own business and was used to doing for others, being the main supporter for our family and those of his employees. He was the one everyone called to ask about their cars (his business), music (his passion) or trivia questions (his phenomenal memory). Being able to always help others gave Sam great satisfaction and really defined who he was. Switching roles to become the one that others helped was an extremely difficult adjustment that took months.

Through the help of a counselor, we both became aware that allowing others to give was a gift in itself. Trying to repay them was disallowing them the ability to be the giver. Let someone help you in and out of the car, let them bring in meals. We all know how good it feels to help others, so learn to accept the help.

Adjusting to changes was, for us, a depressing downslide. Many have years to adjust to using a power chair, traveling in an accessible van or using their brand-new accessible bathroom. Sam's decline was so quick that he only used the bathroom two months before it just was too tiring, the van only has 1,500 miles on it and the power chair just isn't comfortable any more. We went from enjoying the new tools to seeing

them unused. Learning to prioritize what is important is a constantly changing role for each of us.

Guilt — that small word has such a big effect on our lives. Sam constantly was saying "I'm sorry for what you have to go through." I think it was the first time in our 36 years together that he ever felt guilty about anything. I, on the other hand, have grown up feeling guilty about everything. As a caregiver, my guilt has centered on being healthy, being able to walk and go to work, even the smallest things like eating with a fork and scratching my nose. I feel guilty — even though I know I shouldn't — about not being at Sam's side constantly to do what he wants or needs, guilty about going to the park with our grandchildren or visiting them at school.

Denial, frustration and anxiety play huge roles for each of us. I had a lot of frustration when Sam refused to accept losses, such as the ability to work or to use a walker. My role became one of a bad guy. I had to be the one to say "no, you can't do that anymore." I hated being the nag, but it had to be done to assure his safety. It can be difficult to convince a person with ALS that using aids like a power chair and a lift actually increase your independence. It's all part of learning to live with ALS.

For me, the biggest role change was becoming a nurse, in addition to wife, mom and grandmother, and professional. I always said that the last thing I ever wanted to be was a doctor or nurse — I hate needles. But, since December 2006 when Sam got his tracheostomy and went on a ventilator 24/7, I've become an expert at suctioning, changing the trach, understanding the vent's operation, giving medicines and even shots.

Another recent new role for me has been being Sam's voice — to his doctor, nurses, family and friends. For someone who always was talking, telling stories and jokes, this change has been extremely frustrating. I try to convey his feelings and words as best as I can, but it's not always successful. I'd recommend that each family talk early on about the important medical deci-

sions that need to be made, when you're not in crisis, so that everyone is on board with your choices.

Last, don't forget to take care of yourself. If we don't take time to enjoy ourselves — whether it's gardening, exercising, reading, or going out with friends — we'll be run down and feel too much anger at our role as a caregiver. It's a full-time job and everyone needs a break.

My message to everyone is that we can change — we can do what we need to do when crises arise. To answer the question that I hear most often — "How do you do it?" — I say that it's done out of love, that it's my turn to be the caregiver. We don't know what life has in store for us, but we can learn to handle situations with the help of family, friends and the support of the many wonderful charitable organizations. We don't have to deny the anger, exhaustion, fear and all the other feelings that we all feel — but we can deal with them, get beyond them, and not be all-consumed.

(Reprinted from the MDA/ALS Newsmagazine, November/December 2007)

“We were able to remain intimate for a long time. Adjustments must be made, but he told me even the close cuddling, skin to skin, brought him comfort and pleasure. Don't stop until you can't do it anymore — be it eating, walking, being intimate or just living! **”**

“I am not sure why I shy away from making love but I seem totally uninterested. Being intimate brings you closer to someone and maybe subconsciously we are trying to separate ourselves.

If I was to have sex with him it would be so much work. There is no romance because he can't move. I would have to undress him and so on. It would become another chore. **”**

Some spouses find it difficult to maintain a sexual relationship because they see the partner in a clinical way or are too exhausted. In a Catch-22, antidepressants can subdue the libido, as can untreated depression. A partner who only makes demands, never shows thanks or ceases to express affection can put up a barrier to intimacy. Counseling or respite from caregiving may help.

A person with ALS needs to give his or her spouse affection and time to be intimate. If a caregiver provides chores all day, and then is expected to "perform" sexually, it becomes another task, not lovemaking. Both partners can make romantic gestures through words or planning a time for intimacy.

Catheters, vents and immobility make it necessary to adjust sexual positions and actions, but offer a great opportunity for imagination.

One couple took advantage of the times the partner with ALS fell on the floor and was uninjured, and another man says, "Thank God for sturdy ceiling lifts."

Both partners need intimacy and a way to be spouses and lovers first, caregiver and care receiver second.

For more information:

The Ultimate Guide to Sex and Disability: For All of Us Who Live with Disabilities, Chronic Pain, and Illness, by Miriam Kaufman, Cory Silverberg, and Fran Odette, Cleis Press, 2007

In Sickness and in Health, by Gail Lynch, Fairview Press, 2002

Enabling Romance: A Guide to Love, Sex and Relationships for the Disabled (and the People Who Care About Them), by Ken Kroll and Erica Levy Klein, No Limits Communications, 2001

Always On Call: When Illness Turns Families into Caregivers, ed. by Carol Levine, United Hospital Fund of New York, 2000

For Better or For Worse: A Couple's Guide to Dealing with Chronic Illness, by Beverly Kievman with Susie Blackmun, Contemporary Books, 1989

Mainstay: For the Well Spouse of the Chronically Ill, by Maggie Strong, Penguin Books, 1988

Medication

For information on how medications can affect emotions, see "Anxiety," page 100; "Dementia," page 108; and "Depression," page 110.

Parenting

Children of all ages need their mothers and fathers, whether the parent has ALS or not. Parents with ALS will find new ways to interact with their children and remain a central part of their lives. Caregivers can help parents with ALS continue to fulfill their role.

Early in the disease, parents can make the most of the time shared with children and shared physical activities: Now's the time to teach a grandson to fish, take the kids to Disneyland, go sailing or hiking.

When playing ball and gardening become less feasible, caregivers can suggest that the parent with ALS help with homework, read to or with kids, attend games and concerts, or simply talk and listen. They can look up information on the Internet together and share movies and TV. A child may enjoy being the parent's "hands" in the kitchen or in a board game, with the parent directing the child what to do.

A parent with ALS should continue to set rules, make decisions and exercise discipline of children. Kids need to know this person is still the parent, no matter what his or her physical condition.

Parents should encourage children to continue their lives as usual — to study, see friends, participate in sports or other activities, pursue their interests. A child shouldn't feel that his parents have "chosen" caregiving over him, though parents may not be quite as easily available. A helpful relative or neighbor can pitch in to drive kids to practices or other activities.

Invite children to ask questions about ALS so they won't be confused or frightened. One 10-year-old couldn't understand how his grandmother could live without eating. When she showed him her feeding tube and explained how it worked, he was relieved that she wasn't "starving to death."

For the parent with ALS, children are a source of joy and hope. They give something to live for, a reason to fight the disease. Caregivers can help the loved one set goals such

“I am 46 and have a wonderful husband and four beautiful children. I need them as much as they still need me. I am still a wife and mother. They still come to me for advice and homework help. They laugh with me and cry with me. I can't imagine life without them. **”**

“I got to thinking about leaving things for my family when I pass away so that they can remember me when I'm gone. I want my children to remember all of the time our family spent together and all of our favorite memories. I made four scrapbooks for my daughter and four for my son. We're putting together a book specifically dedicated to the family's journey with ALS, with mementos from walk-a-thons and fundraisers, articles we've written for the local newspaper and notes from family members. I've also written journals to my children, telling them stories about things they did, and have made several videotapes as well. Start early before the disease progresses too far. Use your own handwriting in a journal while you still can so your loved ones can have a part of you. **”**

as attending a son or daughter's graduation or wedding; this provides a positive focus and a determination to beat the life expectancy predictions.

Older children may willingly help with caregiving, thus feeling their contributions are important. Early chores could include bringing things to Dad or Mom, tying shoes, clearing the floor so the wheelchair won't be blocked, and simple household tasks like feeding pets. Teens may become adept at handling feeding tubes, ventilators and positioning. Remind a child who's reluctant to help that this is a way to show love for the parent. Some will come around eventually.

Encourage the parent with ALS to think about a legacy to leave for the kids. Caregivers can help the parent create a memory box or scrapbook containing souvenirs, handmade

crafts or photos; write notes or letters to be read on birthdays or wedding days; or record family stories and messages about the parent's hopes and dreams for their children. Stories of their own lives or their family history will be valued by children someday. These tokens, made especially for each child, will remind children long into the future that Mom or Dad loved them and lives in their hearts.

One mother knitted blankets, sweaters, caps and booties for each of her children to use for the grandchildren she would never see. She wanted to express her love for them.

At some point, children will ask, or will need to be told, that the parent with ALS is facing death. The information should be given in age-appropriate language, with reference to the family's spiritual beliefs. Answer questions but don't overload the child with information. Give the child a chance to say goodbye, and be sure he or she understands the parent doesn't want to leave but the illness is making it necessary.

See Chapter 9 for more about children in grief. See Chapter 7 for information on preparing guardianship plans for children.



For more information:

"Having Children After an ALS Diagnosis," MDA/ALS News-magazine, July-August 2010

"Role Reversal: When Children Care for a Parent with ALS," MDA/ALS News-magazine, November-December 2008

"When a Loved One Has ALS: Tips for Helping Children Cope," MDA/ALS News-magazine, March 2008

"Making Memories Last Longer Than a Lifetime," MDA/ALS News-magazine, January 2008

"A Legacy: Sharing Your Life Story," MDA/ALS News-magazine, November 2004

"Parenthood and ALS — Love Wins Out Over Doubts," MDA/ALS News-magazine, June 2002

Grandmother's Memories to Her Grandchild, and Grandfather's Memories to His Grandchild, and similar titles, by Thomas Kinkade, Thomas Nelson, 2004

Adaptive Baby Care Equipment: Guidelines, Prototypes, Resources, by Kris Vensand, Judith Rogers, Christi Tuleja and Anestra DeMoss, Through the Looking Glass, 2000

Bigger Than the Sky: Disabled Women on Parenting, ed. by Michelle Wates and Rowen Jade, Women's Press, 1999

Patient's emotions

A primary caregiver needs to help the person with ALS handle emotions while respecting his or her psychological process.

- Remember that individuals have different timetables for processing grief.
- Encourage the loved one to talk about feelings with the caregiver or other confidant, or at support groups.
- If negative feelings linger, seek out counseling through the MDA clinic's social worker.
- If abuse, anger, anxiety or depression are out of control, get medical help.
- Be aware that refusal to comply with a doctor's instructions or consider assistive technology can sometimes (not always) be due to denial, depression or frontotemporal dementia.

- Be aware that sometimes crying isn't due to sadness but is a disease symptom (see "Pseudobulbar affect," page 54).
- Provide assurance when possible; at other times, just listen and show you understand.
- Remind the person of the many things that still are possible while living with ALS. Talking to a long-term survivor of ALS or someone who's using technology to speak might help.
- Point out that the family is frightened and hurt to see their loved one suffer. They want him or her to stick around as long as possible. Treatments for symptoms or assistive technology may make caregiving easier for them.
- See the clinic physician about depression treatment or to check for signs of dementia.
- Respect the person's decisions about medical and daily care, even if some choices are contrary to the caregiver's.

Peace and priorities

One key to living with ALS is acceptance and restructuring life according to the new rules set by the disease. Some people with ALS and some caregivers find happiness and peace by focusing on personal growth and learning to live in the moment.



After receiving his diagnosis of ALS, Frank Andrews was determined to continue his beloved scuba diving.

“I was ready to rejoin the work force, or so I thought, as I waited for [my husband's] condition to improve or level off. My reality check came the day I walked into the bedroom and saw him using his teeth to pull up the sheet. I was crying on the inside but remained calm on the outside, realizing his frustration and how upsetting this was for both of us. **”**

“I have been able to slow down, look around, and discover the parts of the human experience that are truly fulfilling. Things like my connection with friends and family. These people have always loved me and I them, but I could not understand the depth and importance of these relationships until the polished, hardened, businesslike exterior that I used to occupy was slowly stripped away ... These things do not require a functioning body to enjoy. **”**

Someday is here. It's time to do the things you've dreamed of. Support your loved one in considering new priorities and viewpoints such as:

- Stop worrying about the future. Live in the present.
- Remove the stress of holding a job and meeting deadlines.
- Make every moment meaningful in whatever way they choose.
- Explore their spiritual side.
- Learn and explore intellectually.
- See friends and make new friends.
- Use the computer to learn and stay in touch with people.
- Tutor students or work as a consultant.
- Mend broken relationships.
- Write articles, poetry or books.
- Remain close to children and grandchildren and watch them grow up.

- Enjoy nature or other pleasures.
- Fill the days with things they love: reading, movies, games, laughter, music, pets, conversation.
- Develop new ways to help people with ALS or other disabilities.
- Mentor people with new ALS diagnoses.
- Educate the public about ALS through writing, media interviews or public speaking.

For more information:

Recrafting a Life: Coping with Chronic Illness and Pain, by Charles Johnson, Routledge, 2002

Support

While the primary family caregiver is closest to the person with ALS and may spend most of the day and night with that person, others want to help.

A huge amount of support is available. To reiterate some of the important points made throughout this book:

- Ask the medical care team for help with the loved one's problems or the caregiver's health.
- Get information about the disease and think ahead about equipment and medical decisions.
- Learn everything about assistive equipment and use it.
- Attend support groups or follow online listservs for ALS caregivers. They can provide answers to very specific questions based on personal experience.
- Find an emotional support system that nurtures you, and schedule respite time.
- See an individual counselor or family counselor if problems become overwhelming.
- Use antidepressants, anti-anxiety drugs or sleep aids if they help.
- Cherish your loved one. Use the time to say everything you want to tell them.
- See Chapters 7 and 8 for more specific information about agencies, financial assistance, recruiting

friends and family, and other kinds of help.

- Turn to MDA, its staff, support groups, clinics, publications, videos and other resources.

Sometimes attending a support group is difficult at first; it may bring you and your loved one face to face with people in later stages of ALS, which can be scary. Most people find that the connections, the information and the understanding they derive are well worth any initial discomfort. If an initial support group visit is an overwhelming experience, consider trying again in a few months or a year, when your feelings may have changed.

“A support group is very empowering because you may be upset about something, but somebody there has already been dealing with it. You know that they understand what you are going through, or they usually have some kind of practical advice to make you feel like there is something you can do about it.”

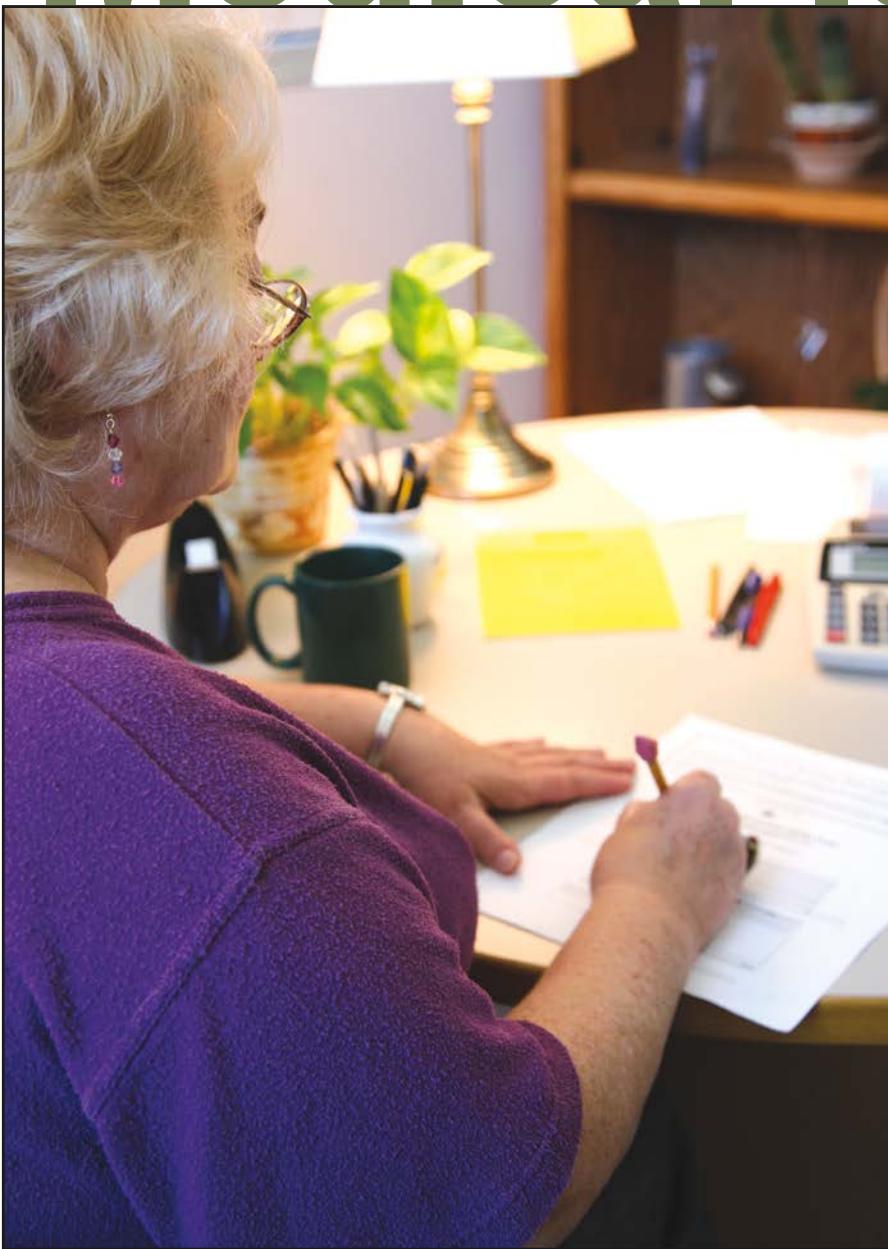
“Socially, it's kind of hard because during the day now, I don't get out much anymore. The group helps with that. We're not just together to share some kind of difficult thing, we're there to have a good time together with people that we've come to like and enjoy.”

“How can you continue to be a good, loving spouse and feel so numb at the same time? Any suggestions for a caregiver who has poured so much out that the cup is drying up?”

“I make a three-hour drive to Houston for the support group. I'm learning what kind of help is out there, like with hospice, and I attend with a different family member each time. Every time we go, we learn something new.”

Chapter 7

Financial, Legal and Medical Issues



Make a Plan

ALS, with all of the equipment, medications, monitoring and daily support required, is expensive. It can easily deplete even a comfortable family's resources. Financial worries often are a major stressor for family caregivers.

During the years in which ALS determines a family's way of life, financial decisions take on new priorities. ALS adds the following to the normal family budget:

- home modifications
- medical bills
- therapies
- durable medical equipment such as wheelchairs
- supplies and maintenance such as wheelchair batteries or liquid nutrition
- home care assistance
- prescription drugs and supplements

Early in the disease, evaluate family income and expenses and project ahead five, then 10 years. Take into account that the person with ALS will likely have to stop working — it's difficult to project when, as each timetable is individual. Often, the primary caregiver gives up his or her job soon after to devote full time to the caregiving role.

This chapter lists a variety of financial options. Many of them take a lot of research and questioning to understand; consider enlisting someone in your family or circle of volunteers (see Chapter 8) to help with this homework. Or, an *elder law attorney* or *benefits counselor* (see sidebar, page 132) could clarify your position with regard to financial resources. At any rate, the more information available, the better prepared a family is to make decisions and seek out financial programs at the most beneficial times.

Finding funds — private sources

Suzanne Mintz, writing in her 2008 book, *A Family Caregiver Speaks Up*, says, "One of the things that frustrates family caregivers more than others is the fact that they are left on their own to wade through a patchwork sea of disparate programs that may not be open to them or meet their needs. Everyone working in the field of caregiving agrees that our system of social supports is hopelessly

“While legal advice is expensive, it can save you a lifetime of aggravation.”

fragmented and insufficient. Family caregivers need the mental acuity and passionate perseverance of a Sherlock Holmes to even solve one part of their support-needs puzzle. Even medical and social service professionals have trouble finding what they need when caregiving turns personal."

That's the unfortunate picture: ALS is hard enough to deal with, but on top of that there's a confused tangle of red tape. The information and resources listed here can help sort out some of that confusion.

In addition to looking into what major government programs and health insurance may provide, the person with ALS should consider these ways to maintain income or acquire other funds (see "Resources" at the end of this section for more details):

- **Continue working part time, from home or as a consultant.** The Americans with Disabilities Act doesn't allow an employer with more than 15 employees to discriminate based on disability. An employer may be willing to modify job duties or conditions as long as an employee still is productive.
- **Look into early retirement.** This allows the employee to receive some income and to retain medical insurance. Under COBRA (the Consolidated Omnibus Budget Reconciliation Act), with proof of disability, a retiree is entitled to continued medical insurance coverage for up to 29 months or until the person qualifies for Medicare — at the retiree's own expense. COBRA also applies to employees who resign or are fired.
- **Exercise economies in everyday life.** Cut unnecessary expenditures and look into less expensive brands of products. Saving a few bucks a week can add up. Trade in the brand new car for one with a few years on it. Sometimes you can take the car back to the dealership and explain the situation, and the dealer or manufacturer will cancel the contract or lease. Buy in bulk or ask a friend to split a large order or pick up a sale item. Investigate lower-priced cable TV and cell phone

service; tighten up little budgetary “leaks” where small amounts of money are being spent unnoticed.

- **Take advantage of tax deductions related to ALS.** As of 2007, if unreimbursed medical expenses, including transportation to doctor visits, exceed 7.5 percent of adjusted gross income, you qualify for deductions on Form 1040, Schedule A. (This percentage goes from 7.5 to 10 percent in 2013.)

Also deductible are “impairment-related work expenses,” which are disability adaptations and services that enable a person to continue working. This can include special computer programs and technological adaptations, hired work assistants and, sometimes, personal assistance at home for getting dressed and ready for work. The standards for claiming impairment-related work costs are less stringent than for medical deductions. If the device or service is “helpful and appropriate,” the deduction often qualifies. Keep receipts!

People with disabilities may, under certain circumstances, receive a tax credit for child and dependent care expenses or qualify for the Credit for the Elderly or Disabled.

By the way, if the person with ALS is no longer able to sign his or her name, the caregiver will have to do that. For official documents such as tax returns, you’ll need to show a power of attorney form that gives you authority to sign for your loved one.

- **Get a reverse mortgage.** People over 62 who own all or most of their home can borrow against that equity without having to pay it back. The homeowner retains title and pays property taxes, insurance, maintenance

“Keep all papers on out-of-pocket expenses. This includes home improvements, showers, special toilets, ramps, hand rails, lifts on vans, etc. All this is deductible for IRS and state tax over a certain amount. So keep that shoebox handy to keep your records of what you pay out. We did this for 20 years with my husband’s care and it never was questioned by IRS or the state.”

costs, low interest and some high upfront loan fees. The loan — from government or private sources — doesn’t have to be repaid until the last surviving owner vacates the home, at which point the home is sold to pay the bill.

- **Borrow to modify the house.** Those who need to make physical adaptations to their homes because of disability may be eligible for a *Title I Home Improvement Loan* insured by the U.S. Department of Housing and Urban Development (HUD). The loan can be used to remove architectural barriers, hazards or inconvenient features in the home.

A home equity loan (second mortgage) can serve this purpose and also be used for ordinary expenses. Some families advise getting an equity line of credit or home equity loan for the maximum amount, just to have funds available. This provides funds but, of course, must be repaid. It’s best to apply for these loans while at least one spouse is still employed full time.

- **Let friends pitch in.** Friends, neighbors, co-workers, church members or civic club members who want to help someone with ALS can be put to work organizing a yard sale, concert, spaghetti dinner or other event to help cover a specific expense such as a wheelchair-accessible van or home modification.
- **Raid retirement savings.** Money put into an IRA, 401k or 403b plan, or other retirement account normally can’t be touched until the saver is 59.5 years old. People younger than that who need cash for ALS expenses may be able to borrow or take cash from the account. Often there’s a stiff financial penalty involved, but some plans make an exception for disability-related expenses.



- **Use disability insurance.** When unable to work, people with long-term disability insurance receive an income for life or for a specified time. The amount is usually a percentage of the person's previous salary. Some employers provide disability insurance as part of a company benefits package.

Be aware that some long-term disability policies will reduce the benefit if the person with ALS, his or her spouse, or children are receiving other benefits such as Social Security. *Read the fine print!*

- **Move assets.** In anticipation of eventually needing Medicaid or Social Security Disability Insurance benefits (see page 133), diminish the assets of the person with ALS. They can be moved into a spouse's name alone. Or, they can be distributed to heirs up to a certain limit per year. This is tricky. Be sure to check with an *elder law attorney* first. An elder law attorney specializes in issues related to the elderly or people with disabilities.

- **Use life insurance.** There are several ways in which a person with ALS may use life insurance settlements to help with expenses, including borrowing against the policy: Many insurers allow people to take up to 50 percent or so of the benefits upon confirmation of a terminal illness, and the proceeds aren't taxable. Premiums must continue to be paid, and the beneficiary will only receive what's left after the loan is repaid.

Some life insurance policies have ***disability waivers***, which suspend payment of premiums as long as the person is disabled but keep the policy in force. That policy can be used as collateral in a loan application.

Another choice is the ***cash surrender value***, in which the insured person terminates the policy and receives a cash settlement. However, this amount is much less than the full amount a beneficiary would receive. It's also possible to sell a life insurance policy as a ***viatical settlement***. Investors purchase the policy at less than the death benefit.

Taking cash from a life insurance policy before death will lessen its eventual value; decide whether that loss to the beneficiary is a good tradeoff for cash to help cover ALS expenses.

- **Check with your Area Agency on Aging** or state department of social services to see whether your

loved one can hire a family member as a caregiver and receive payment for his or her services.

Consult an elder law attorney or a trusted financial adviser acquainted with disability issues before making any major financial decisions.

Resources

Finding funds — private sources

"It's All in the Plan — How to Start Long-Term Financial Planning," MDA/ALS Newsmagazine, October 2007

Financial Aid for the Disabled and Their Families, by Gail Ann Schlachter and R. David Weber, Reference Service Press, 2006

Plan While You Can: Legal Solutions for Facing Disability, by Roger W. Anderson, 1stBooks, 2003

The Cost of Caring: Money Skills for Caregivers, by Anne M. Johnson and Ruth Rejnis, Wiley, 1998

Information and Referral
211.org

Modest Needs
(212) 463-7042
modestneeds.org

Offers self-sufficiency, back to work, and independent living/quality of life grants.

Ride for Life
(631) 444-1292
rideforlife.com
Offers respite grants and legal grants for people with ALS.

Working

A Consumer Guide to Handling Disputes with Your Employer or Private Health Plan, by the Kaiser Family Foundation and the Center for Consumers Health Choices at Consumers Union, 2005

Benefits Management for Working People with Disabilities: An Advocate's Manual, by Edwin J. Lopez-Soto, Esq., and James R. Sheldon Jr., Esq., Greater Upstate Law Project, 2002

Benefits Counselors: Who Are They and Do I Need One?

by Sandra Fusion

There are so many benefits, both financial and assistance-oriented, to which individuals are entitled. These include community-based programs, state or federal assistance programs, as well as retirement and insurance programs.

One of the issues with having so many benefits available is that it's difficult to track them and find out which ones are appropriate for the situation. This is where a benefits counselor can help. The term "benefits counselor" can be applied to either a paid staff person for an agency or a trained volunteer. In simple terms, a benefits counselor is someone who reviews existing information about your financial situation and makes suggestions about benefits for which someone may be eligible, or managing existing benefits.

Who are benefits counselors?

Generally, benefits counselors work with individuals age 60 and older. If someone has a disability, they also are entitled to receive benefits counseling information. Many work with or volunteer for agencies like the Area Agencies on Aging (AAA). With the AAA, there are stringent criteria for benefits counselors and certification levels are available. The various certification levels designate the types of cases for which someone can receive assistance.

Individuals receive free assistance from a benefits counselor. It is not attorney representation, however. Individuals who need legal representation still may receive a benefits counselor; however, the counselor will not be able to represent them in court.

How can they help?

It can be confusing to figure out eligibility requirements, how to apply, what types of documentation are needed and many other issues that arise when looking at benefit programs. Throw in supplemental insurance policies, and you have a recipe for confusion.

Trained benefits counselors can sort through the "confusion" to help develop a cohesive benefits plan. The benefits counselor serves a valuable role in discovering possible avenues for benefits that give families alternatives that may not have been known before.

The types of information that benefits counselors can examine include (but may not be limited to): insurance benefits; Social Security benefits; Social Security Disability benefits; Medicare (including the new prescription benefit section, Part D); prescription assistance programs (for individuals not eligible for Medicare); income tax benefits and credits; retirement benefits; veterans benefits; community-based benefits; state program benefits, including home care alternatives; advocacy with agencies, if needed; and referrals to community-based organizations or government agencies.

Make sure you have documentation available when you speak to a benefits counselor. Things to consider taking with you include: recent statements from your insurance company; recent bank account statements; medications you are currently taking (for Medicare Rx or other prescription benefit programs); retirement statements concerning benefits you are already receiving; Social Security statements and card (if available); disability benefits currently receiving; and any other state, federal or community program where you are currently enrolled.

What if you've applied for services and have been denied, yet you still feel that you qualify? A benefits counselor can examine your case individually and try to advocate on your behalf. Advocacy does not guarantee services, however.

Advocates may be able to sort through the requirements and find out if there has been miscommunication, missing documentation, or other communication barriers that prevent you from receiving specific benefits. If you still are denied benefits, at least you will feel like you have received the total attention of the "system."

Where do they work?

You can search for a certified benefits counselor through local Area Agencies on Aging (AAAs). Other organizations include your local human service offices, county welfare offices, and community-based organizations that serve the elderly and/or disabled.

Another method to find a benefits counselor is by calling an Information and Referral helpline. In more than 46 percent of the United States, you can dial 2-1-1 and reach a trained professional who can identify organizations in your community where benefits counselors work. If your area does not have access to 2-1-1, usually there is one point of entry into the human service system. Some places call it a helpline, while others call it information and referral. To find out if your community has access to 2-1-1, you can look online at 211.org. The nationwide status map also can give insight into where to call if your area is not served by 2-1-1.

Learning about available benefits can be challenging. It's important to have a trained professional review your situation and point out avenues you may not have considered investigating.

(Excerpted with permission from "Today's Caregiver," caregiver.com)

Retirement

COBRA (Consolidated Omnibus Budget Reconciliation Act)
(866) 4-USA-DOL

www.dol.gov/ebsa/cobra

Federal law provides certain former employees, retirees, spouses, former spouses, and dependent children the right to temporary continuation of health coverage at group rates.

Tax deductions

"Timely Tips To Help Limit Your Tax Liability," MDA/ALS Newsmagazine, January 2008

"ALS Expenses Can Be Tax Deductible," MDA/ALS Newsmagazine, January 2004

Internal Revenue Service

(800) 829-1040

irs.gov

Publication 501: Exemptions, Standard Deduction, and Filing Information

Publication 502: Medical and Dental Expenses

Publication 503: Child and Dependent Care Expenses

Publication 524: Credit for the Elderly or Disabled

Publication 525: Impairment-Related Work Expenses

Publication 529: Miscellaneous Deductions

Publication 596: Earned Income Credit

Publication 907: Tax Highlights for Persons with Disabilities

Publication 910: Guide to Free Tax Services

Publication 3966: Living and Working with Disabilities/Tax Benefits and Credits

Reverse mortgage

"Reverse Mortgages: Money from Home," MDA/ALS Newsmagazine, February 2006

AARP

(800) 209-8085

aarp.org/money/revmort

National Center for Home Equity Conversion
reverse.org

National Reverse Mortgage Lenders Association

(866) 264-4466

reversemortgage.org

U.S. Department of Housing and Urban Development

(800) 569-4287

hud.gov

Search "reverse mortgage" for selection of information.

Home modification loans

Section 8 Made Simple: Using the Housing Choice Voucher Program to Assist People with Disabilities,
by Ann O'Hara and Emily Cooper, Technical Assistance Collaborative, 2003

"Affording Accessibility: Home Modifications"

United Spinal Association

(718) 803-3782

unitedspinal.org

Life insurance

American Council of Life Insurers

(202) 624-2000

acli.com

Viatical and Life Settlements

viatical-expert.net

Vitatic Association of America
(202) 367-1136

Also check with your state's attorney general, office of consumer protection, insurance commissioner or department of insurance.

Finding funds — government sources

People with ALS and their caregivers have paid taxes to the state and federal governments for many years, and an ALS diagnosis means it's time to claim some of the benefits those taxes support. First, the person with ALS must choose between continuing to earn an income and taking advantage of government programs, which define disability as inability to work.

Some federal programs require that applicants not earn any income, and in some cases, have only minimal assets. See "Resources" at the end of this section, for where to apply and where to read about these programs in detail.

Many federally funded programs are directed at the state level, and each state's program is different. Some state programs are more advanced than the federal legislation, but many are below federal guidelines. The information here is general. To find out what public programs exist in your state, go to caregiver.org and click on "Family Care Navigator."

NOTE: Social Security, Medicare and other government programs described here may be changed by Congress at any time. Consult an adviser or the offices of these programs for the latest guidelines. Don't sign up for any program before understanding the long-term implications. Other families in your MDA support group may be able to recommend a good adviser.

Social Security Disability Insurance (SSDI)

To qualify for this program administered by the Social Security Administration, a person must have a disabling condition that prevents working, and the condition should be expected to last at least 12 months. The person must be under 65 and have paid a qualifying amount into the Social Security system throughout the working years.

People with ALS who meet the qualifications are granted "presumptive" or automatic eligibility for SSDI benefits,

“ You can benefit from Medicare for hospice services, and payment for rental equipment, medications for pain and symptom control, visits by a registered nurse, home health aide, social worker, volunteers, etc., indefinitely as long as the person continues to decline over time. Not only does it make good sense financially, but also in terms of support for you and your family. **”**

meaning they should need nothing more than a doctor's statement of diagnosis to prove qualification. The person will receive a monthly check indefinitely, or until able to work again. At age 65, disability benefits turn into Social Security retirement benefits. SSDI is available without regard to family assets or income.

Sign up for SSDI at any Social Security office as soon as the diagnosis of ALS is made or as soon as the person stops working. Benefits won't begin until five months after disability status has been granted, and the approval process may take time. Bring employment and medical information, including W-2 forms, copies of doctors' diagnoses, and their addresses and phone numbers for verification. It's possible to apply for SSDI online or by phone. If the person you talk to doesn't seem to understand or to be knowledgeable, go to the local Social Security office, or consult another adviser.

SSDI recipients with ALS can begin receiving Medicare Part A (with Part B optional (see right) as soon as they get SSDI.

Supplemental Security Income (SSI)

SSI is a monthly benefit paid to adults of any age who are aged, disabled or blind and who have limited income and resources. People with ALS who have never been employed may be entitled to SSI. When applying, bring Social Security number, birth certificate or proof of age, as well as information about the diagnosis and your home and medical care.

People who receive SSI benefits are often eligible for food stamps and Medicaid too.

Social security income for children

Benefits are paid to minor children when a parent who is eligible for Social Security becomes disabled or dies. The benefit depends on other family income; the child must be under 18 and unmarried; benefits may continue past age 18 for full-time students or those with disabilities.

NOTE: Some long-term care insurance policies state that Social Security or other income paid to youngsters can be subtracted from the income paid by the insurance to the parent.

Medicare

Part A

Medicare refers to health insurance administered by the federal government; it's available to people over 65, or those under 65 with disabilities. A person with ALS qualifies for Medicare as soon as SSDI benefits begin.

The Medicare beneficiary pays no premiums for Part A, which helps cover:

- care in the hospital
- related services after leaving the hospital
- partial coverage of post-hospitalization hospice services
- respite care for caregivers of those in hospice
- medically necessary skilled nursing care at home or in a care facility

Certain deductibles and coinsurance amounts apply. It's possible to buy into Part A if you're over 65 but never paid Medicare taxes.

Part B

While Medicare Part A costs the consumer nothing, its coverage is generally limited to medically necessary hospital or hospice care. Many more services important to people with ALS are available under Medicare Part B, which requires payment of a monthly premium. Part B helps pay for:

- doctor visits
- outpatient care
- preventive care and diagnostic tests
- ambulance



- emergency room services
- lab services
- physical, occupational or speech-language therapy
- outpatient mental health care
- custodial care in nursing home
- canes and crutches
- commode chairs
- durable medical equipment
- orthotics
- walker or wheelchair

Parts A and B both pay toward home health care under certain circumstances. For details, go to medicare.gov/publications and enter "10116" into the search box.

Medicare Parts A and B generally pay 80 percent for health care and equipment, so most people also carry a Medigap or supplemental policy to pick up the rest (see page 140). For conditions like ALS, Medicare coverage for home care, therapies and nursing homes is limited to a short period of time and skilled nursing care only. To get coverage for the ongoing services that people with ALS require, a doctor's letter of medical necessity (see page 140) may have an impact.

Insurance systems are based on an "acute care" model, geared to the needs of those with a temporary illness or injury who are expected to improve. With either Medicare or private health insurance, it can be difficult to get care for a chronic condition such as ALS. However, families

who've dealt with this situation have found a plethora of solutions (see page 140).

Part C

Medicare has introduced Advantage Plans which allow a choice of HMOs, PPOs or other plans. These include Parts A and B of Medicare, and Part D is optional.

Part D

This newest Medicare benefit, which went into effect in 2006, is prescription drug coverage. It requires a separate application and enrollment, and sometimes charges a premium.

Programs for Medicare Beneficiaries with Low Incomes

Each state is required to offer a Qualified Medicare Beneficiary Program and a Specified Low-Income Medicare Beneficiary Program to help Medicare recipients with low incomes; these may help pay additional health care expenses.

Medigap insurance

This term refers to private insurance policies that can supplement the coverage provided by Medicare.

“Every two years I had to fight our insurance all over again. I did my homework, research and wrote the most factual letters with the laws for disabled people, the most compelling letters I could, and sent them to everyone that had anything to do with my husband's case. Our doctor called our insurance company and wrote letters and threatened to take it to the newspapers.

Every year, like a Cecil B. DeMille drama, the insurance would take it to the 12th hour and then inform us, 'OK, for another two years your husband will get 16 hours of in-home RN or LVN knowledgeable care.'

We were so grateful to the wonderful nurses that came into our lives for seven years. I am most grateful to the two liaisons that fought for [my husband] and also grateful to our doctor. **”**

Medicaid

Medicaid, also known as the Medical Assistance Program, is a federal program that's administered separately by each state, that helps cover medical costs of people with low incomes. Those with higher incomes may be eligible if their medical expenses exceed a given percentage of their annual income. Medicaid benefits vary from state to state, but coverage for long-term care usually is more extensive than that provided by Medicare. Medicaid may cover in-home and nursing home care for an unlimited time period, and includes nonmedical as well as medical care. Medicaid guidelines and eligibility requirements vary from state to state.

Eligibility is based on three criteria: care needs, income and assets. Earned wages, Social Security, disability insurance, retirement, pension and alimony all count toward the income cap for Medicaid eligibility. Assets include bank accounts, brokerage accounts, stocks and bonds, loans, annuities, and any resource of value owned jointly by applicant and spouse. To learn the rules of your state's Medicaid program, visit medicaid.gov and select your state; or call your Medicaid agency. You also can call the Centers for Medicare & Medicaid Services (CMS) at (877) 267-2323.

Once you're eligible for Medicaid, many states also cover physician, psychologist, rehabilitation, chiropractic, dental, laboratory and X-ray services, hospital visits, prescription drugs and prosthetic devices.

When Medicaid initially assesses a person's assets to check for qualification, that "snapshot" determines how much can be retained by the unaffected spouse. An expert can advise on the details in your state and what assets are counted in the calculation. *Do not transfer assets to other family members before consulting an attorney or benefits expert, as there are very strict guidelines for this process.*

Qualified Income Trust

There are a couple of ways to get around Medicaid's income and asset limitations.

A Qualified Income Trust, also known as a *Miller Trust*, is an account in which you deposit enough income — earned, pension, Social Security, etc. — to reduce your qualifying income for Medicaid coverage of nursing home placement.

Placing funds into a QIT lets you qualify for Medicaid and receive benefits such as long-term care programs. The funds left in trust may be available to leave to the survivor.

Cash and counseling

Another Medicaid program, called Cash and Counseling, gives cash instead of standard agency services to eligible individuals. The beneficiary then can decide how to spend the cash to cover disability-related expenses. It can be used to buy equipment, or pay for home care, or whatever makes the most sense for the individual. Beneficiaries receive counseling to determine the most effective use of their cash; hence the name of the program. At this writing, the program only is available in about 15 states on a trial basis, but soon may be expanded.

Medicaid state waivers

Many states have waivers to the Medicaid program that cover services over and above regular Medicaid benefits. They may cover specific needs or provide enough care to allow the person to remain at home, or even pay family members to be caregivers.

Other government funding sources

Alternative Financing Program

This federal/state program grants low-interest loans to people with disabilities, their relatives or advocates in order to purchase assistive technology or services. People who don't qualify for traditional bank loans may find AFPs more receptive to their applications.

Family and Medical Leave Act

Caregivers and people with ALS can take advantage of the Family and Medical Leave Act of 1993 to hold onto their jobs. An individual may be entitled to take up to 12 weeks of unpaid leave to care for a close family member without fear of losing his or her job.

Veterans Administration

ALS researchers have established that veterans of the Persian Gulf War in the early 1990s have a slightly higher risk of developing ALS when compared to other Americans of the same age. It's one aspect of the

phenomenon known as **Gulf War syndrome** (veterans of the Iraq War haven't yet been studied for this effect). The suspected reasons involve environmental exposure interacting with particular genetic factors; the exact mechanism is part of the mystery still surrounding the causes of ALS.

ALS developed as part of the Gulf War syndrome isn't clinically any different from other types of ALS. The Veterans Administration offers a Gulf War registry examination and other benefits for veterans.

If your loved one with ALS is a military veteran, he or she should check with the Department of Veterans Affairs to find out about programs available to veterans with disabilities. Programs may provide those qualified with outpatient medical services, in-home respite care, durable medical equipment, day services for vets and caregivers, lower-cost prescriptions and special assistance for veterans of the Gulf War whose ALS may be service-related.

Independent living centers

Independent living centers (ILCs or CILs) exist in every sizeable city. They focus on enabling people with disabilities to live independently in the community and at home, and the staff usually knows about all types of resources for people with disabilities.

Ask your local MDA office for information about these programs, or visit the National Council on Independent Living site, ncil.org. For a list of centers, see www.virtualcil.net.

P&A and CAP

The **Protection & Advocacy System (P&A)** and the **Client Assistance Program (CAP)** are mandated by the federal government to help protect the legal rights of people with disabilities. P&A and CAP services vary from state to state. To find local offices, go to the National Disability Rights Network website, napas.org, and click on "Get Help in Your State."

Resources

Finding funds — government sources

National Council on Aging BenefitsCheckUp
(202) 479-1200
benefitscheckup.org
Links to various federal and state benefit programs.

SSDI

“Applying for Social Security Benefits Now a Little Easier for People with ALS,” MDA/ALS Newsmagazine, June 2004

“Social Security Changes Rules to Benefit Applicants with ALS,” MDA/ALS Newsmagazine, September 2003

Nolo's Guide to Social Security Disability: Getting & Keeping Your Benefits, by David A. Morton III, M.D., Nolo Press, 2006

Social Security Disability Insurance (SSDI)
(800) 772-1213
socialsecurity.gov or ssa.gov

SSI

Supplemental Security Income (SSI)
(800) 772-1213
socialsecurity.gov/ssi

Social Security income for children

Supplemental Security Income for Children
(800) 772-1213
socialsecurity.gov/ssi/text-child-ussi.htm

Medicare

“An Endless Fight for Essential Benefits,” MDA/ALS Newsmagazine, October-November 2005

“Medicare Covers Cough Device,” MDA/ALS Newsmagazine, September 2003

Center for Medicare Advocacy
“Medicare Basics: A Guide for Caregivers”
(202) 293-5760
medicareadvocacy.org

Centers for Medicare & Medicaid Services
(800) 633-4227
www.cms.hhs.gov

Medicare
(800) 633-4227
medicare.gov

Medicare Rights Center
(800) 333-4114
medicarerights.org

State Health Insurance Assistance Program
shiptalk.org
Offers free one-on-one counseling and assistance to people with Medicare and their families.

Part D

Medicare Part D Appeals: An Advocate's Manual
medicarerights.org

Medicare Rx-Connect
maprx.info

Medicaid

“Medicaid Planning: Look Before You Leap” MDA/ALS Newsmagazine, November/December 2007

Centers for Medicare & Medicaid Services
(877) 267-2323
www.cms.hhs.gov

Medicaid
(800) 633-4227
www.cms.hhs.gov
Or contact your state's Medicaid office or department of health services.

Qualified Income Trust

“A Qualified Income Trust: Keep Your Medicaid Eligibility,” MDA/ALS Newsmagazine, April 2005

National Academy of Elder Law Attorneys
(520) 881-4005
naela.org

Cash and counseling

“New Medicaid Program Gives Recipients Cash, Not Predetermined Services,” MDA/ALS Newsmagazine, December 2003

Cash & Counseling
(617) 552-2809
cashandcounseling.org

Other government funding sources

Aging & Disability Resource Center
adrc-tae.org

DisabilityInfo.gov
(800) 333-4636

Indian Health Services
ihs.gov/medicalprograms/eldercare
olderindians.org

Alternative Financing Program

“Paying for Assistive Technology: Make Alternative Financing Work for You,” MDA/ALS Newsmagazine, January 2007

“Funding Freedom: Tech Loans Provide Alternate Financing,” Quest, May/June 2006

RESNA (Rehabilitative Engineering and Assistive Technology Society of North America) Alternative Financing Technical Assistance Project
(703) 524-6686
resnaprojects.org/afp
National AT Technical Assistance Partnership
resnaprojects.org/statewide

Family and Medical Leave Act

U.S. Department of Labor
dol.gov/whd/fmla

Veterans Administration

“Report Concludes Military Tie Deserves Further Study,”
MDA/ALS Newsmagazine, January 2007

“‘Detox’ Enzyme DNA Links Genes,” MDA/ALS News-magazine, August 2006

“Gulf War Report Available Online,” MDA/ALS News-magazine, January 2005

GulfLink Medical Information
www.gulflink.osd.mil/vet_help/medical.jsp

National Gulf War Resource Center
(866) 531-7183
ngwrc.org

U.S. Department of Defense
www1.va.gov/gulfwar
Gulf War Veterans Helpline (800) 749-8387
Gulf War Veterans Hotline (800) 796-9699

U.S. Department of Veterans Affairs
Registry of military veterans with ALS
(877) 342-5257
durham.hsrdr.research.va.gov/alsregistry.asp

Office of Geriatrics and Extended Care
(877) 222-8387
va.gov/geriatrics

Paralyzed Veterans of America
(800) 424-8200
pva.org

Independent living centers

Independent Living Research Utilization
Directory of Centers
(713) 520-0232
ilru.org/html/publications/directory

Independent Living USA
ilusa.com

Protection & Advocacy

Center for Patient Partnerships
(608) 265-2627
patientpartnerships.org

National Disability Rights Network
napas.org

Help with health care and equipment costs

Private and government insurance plans cover a great deal of the medical expenses associated with ALS, but not everything. Become familiar with which policies cover what, and be prepared to appeal denials of coverage. For more information, see “Resources” at the end of this section.

Private medical insurance

Under federal regulations, an employer's health care plan must include arrangements for continuing an employee's coverage for at least some period of time after the employee leaves the job for reasons including disability. (See “COBRA,” page 133.) The employer also may maintain short-term and long-term disability plans for employees. Check with the company's human resources department.

Most group and private health insurance programs now are offered through health maintenance organizations (HMOs), preferred provider organizations (PPO) or some other type of managed care. Because managed care companies are committed to saving costs, they may present obstacles to complete care for people with serious conditions like ALS. They may be reluctant to refer a patient to a specialist outside the organization, unwilling to cover long-term therapies or deny other claims. All insurance organizations have appeals procedures. Ask for the company's patient advocate, or your employer's human resources department, in order to find out more about these. See “Coverage Denial,” right, for more ideas on dealing with denials.

Medigap or supplemental insurance

It's important to hold onto group coverage through an employer to cover the things Medicare won't cover; if the group policy expires, look into a Medigap or supplemental insurance policy. These can be purchased even after an ALS diagnosis. These policies don't cover as many expenses as long-term care or private health insurance, but they'll help.

If the person with ALS has no prescription drug coverage, and isn't enrolled in Medicare Part D, a local pharmacist should be able to explain which supplemental insurance would cover necessary drugs.



Long-term care insurance

This type of insurance policy covers medical and care expenses at home or in a nursing facility (see page 165).

It's a good idea for a primary family caregiver to purchase this type of policy for himself or herself. The spouse or adult child with ALS isn't going to be able to provide care if the caregiver becomes ill.

Coverage denial

Even under the best insurance plans, specific claims sometimes are denied. When consumers feel the denial violates the policy, they can appeal the decision, often with good results. Some observers say that insurance companies deny a certain number of claims in the belief that people won't bother to appeal.

Some tips for appealing a denial:

1. Get help. The doctor, hospital business office or employee benefits office may be willing to argue that an expense is medically necessary, and this can be a lot more powerful than statements from an individual policyholder. Health care staff at MDA clinics are used to providing these *letters of medical necessity* and usually know the right words for requesting coverage or appealing a denial.

When the insurance denial says that some service, such as physical therapy, is only covered for a certain number of weeks, an effective letter from a physician or specialized therapist might point out some slight change that creates a new or ongoing need for the coverage: new caregivers who need training in transfers, ROM exercises, or positioning in bed and wheelchair; indications that therapy will provide increased functional independence, more safety in transfers and mobility, safer swallowing, improved speech,

etc. A change in condition — the patient is much weaker and is having increased joint pain — can justify sending a PT to the home to instruct caregivers in more appropriate ROM and stretching exercises.

2. Be persistent. It may be necessary to go through several levels of appeals, but showing that you aren't giving up can persuade the insurance company to settle the claim. Ask the insurance company to assign you a case manager.

3. Use the right words. Be sure to cite a specific *medical* problem that the service addresses; stay away from terms like "cosmetic" or "self-esteem." *Never* make the argument that a patient benefit will help the caregiver provide better care.

4. Ask your doctor to try again. Be sure that the letter of medical necessity states each specific medical problem for which the procedure or drug is needed.

5. Consult a lawyer. Many times the threat of a lawsuit (with lawyers copied on letters) gets the insurer's attention and reverses a denial. Use an attorney who specializes in elder or disability law.

6. Use state resources. Consult the state department of insurance, Protection & Advocacy System and Local Agencies on Aging. Write your federal representatives and governor.

Hospice

Hospice care (which can take place in your home or in a facility) focuses on preserving quality of life for a terminally ill person, and provides caring services for the whole family. Hospice services can provide medical equipment, in-home nursing care, social services, respite care, bereavement counseling and other services. A doctor must refer a patient for hospice care. It can be a great source of help for caregivers. (See "Hospice," page 165.)

Hospice care is usually covered by Medicaid, private insurance and Medicare. Most hospice programs require a doctor's statement that a person has six months or less to live, although that statement is just a "guesstimate," not a fact, and people may live for years after having been referred to hospice. It's possible to go on and off hospice several times, if the person's condition changes. To find out more, contact a local hospice organization. Staff can visit your home and do an intake application to see what help they can give.

Prescription drugs and supplements

Several sources of help exist for purchasing prescribed drugs and nutritional supplements. See "Resources" at the end of this section for details on:

- Discount cards and free drug programs
- Buying online from U.S. pharmacies
- Buying and ordering online from Canada
- Medicare Part D
- Suggestions from local pharmacists

Low-cost sources for equipment

Most durable medical equipment, such as wheelchairs and ventilators, is prescribed by doctors, and therapists consult on the proper fit and model. These usually are covered by insurance, often with a deductible paid by the consumer. Using a borrowed or secondhand item — always in consultation with a physical therapist — can save some cash. Most insurance companies will pay for only one wheelchair every five years. Save this benefit for the sophisticated, expensive power wheelchair needed in the later stages of the disease.

Consult with a therapist before buying any mobility equipment on your own, especially something like a scooter that requires some strength to operate, because ALS only may allow a few months of usage before weakness makes the equipment unusable. Other equipment, such as hospital beds, bath chairs or vehicles that accommodate wheelchairs, doesn't have to be so carefully matched to the individual's physical abilities.



There are many ways to find used or temporary equipment, and to lower costs. Here are a few:

- MDA's equipment program has many donated pieces to lend.
 - MDA provides financial assistance with repairs and modifications to all types of durable medical equipment.
 - Rather than going through a local dealership for a wheelchair, CoughAssist or other device, go directly to the manufacturer. It's possible to negotiate a discount, and most companies will handle billing the insurance company or Medicare. Local providers will come to the home to set up equipment such as ventilators and monitor them regularly.
 - Study the health insurance policy or Medicare guidelines, and be sure to choose a device that fits their coverage criteria, or prepare to argue for the one that's needed. Be aware that most insurers will buy a wheelchair only once every five years or so. Don't waste this benefit on a low-cost scooter or used manual chair. Save it for a good power chair with all the bells and whistles.
 - Friends, relatives, church members or support group members sometimes have items to discard or sell.
 - Check MDA newsletters and notices at clinics, and ask MDA health care service coordinators about vendors who have good-quality secondhand equipment. Also ask occupational, physical, respiratory and speech therapists.
 - Look at "for sale" ads on the bulletin boards, websites and newsletters of rehabilitation hospitals, independent living centers, state Rehabilitation Departments and other local disability organizations. Try craigslist.org.
 - Check ads in the classified pages of the daily newspapers or local "shoppers," or even on supermarket bulletin boards.
 - Pawn shops and secondhand stores sometimes have walkers, wheelchairs, hospital beds and the like.
 - Search the Internet under "durable medical equipment" or "wheelchairs"; many websites, including eBay, list and picture secondhand canes, cushions, wheelchairs, vehicles, walkers and other devices.
- See "Resources" for information about resources for buying and selling adapted vans. Check National Mobility Equipment Dealers Association (NMEDA) for reputable sources of vehicles.
 - The Technical Assistance Project maintains local centers that offer assistance in obtaining funding for technology devices; many have equipment lending, exchange or recycling programs. The project is supported by the federal Technology-Related Assistance for Individuals with Disabilities Act funded through the National Institute on Disability and Rehabilitation Research.
 - Watch articles in MDA's Quest and MDA/ALS News-magazine to compare products and get other ideas on cost savings. Ads in Quest sometimes offer a discount for Quest readers.
 - The Alternative Financing Program grants low-interest loans for purchase of assistive technology or services.

Vehicles

See "Resources" for information on ways to finance an adapted van.

Resources

Help with health care and equipment costs

Private medical insurance

Health Insurance Manual: Options for People with a Chronic Disease or Disability, by Dorothy E. Northrop and Stephen Cooper, Demos Medical Publishing, 2007

Insurance Solutions: Plan Well, Live Better: A Workbook for People with a Chronic Disease or Disability, by Laura D. Cooper, Esq., Demos Medical Publishing, 2002

For information on assistance in acquiring health insurance or prescription meds:

County or State Department of Health and Human Services

Social service agencies, including Association of Jewish Families and Catholic Charities.

National Council on Aging
BenefitsCheckUp
benefitscheckup.org
Helps those over 55 find federal, state, and local public and private programs that may pay for some of their medical care and/or prescription costs. A companion site, benefitscheckuprx.org, provides information about prescription medical programs.

HealthInsurance.com
(800) 644-3491
healthinsurance.com
A resource for consumers and small businesses seeking affordable health insurance.

Medigap and supplemental insurance

“It’s All in the Plan — How to Start Long-Term Financial Planning,” MDA/ALS Newsmagazine, October 2007

Long-Term Care: How to Plan & Pay for It, by Joseph L. Matthews, attorney, Nolo, 2004

Offset of long-term care insurance benefits by government assistance to the family. See:
willymsrant.blogspot.com/2006/11/social-security-offset.html
erisaontheweb.com
disabilityinsuranceforums.com

Coverage denial

“Insurance Claims: Don’t Take ‘No’ for an Answer,” MDA/ALS Newsmagazine, April 2006

A Consumer Guide to Handling Disputes with Your Employer or Private Health Plan, by Kaiser Family Foundation and the Center for Consumers Health Choices at Consumers Union, (650) 854-9400, kff.org

Center for Patient Partnerships
(608) 265-6267
patientpartnerships.org

“Insurance Ombudsman”
money-zine.com

Legal Aid Society
Look in local phone book.

National Patient Advocate Foundation
(202) 347-8009
npaf.org
Patient Advocate Foundation
(800) 532-5274
patientadvocate.org

Hospice

See Chapter 8, “Resources,” page 190.

“Not Gloom and Doom: Demystifying Hospice,” MDA/ALS Newsmagazine, January-February 2010

Prescription drugs and supplements

“Playing the Money Game,” Quest, May-June 2008

“Shopping for Health: Is There a Pill to Ease the Pain of High Drug Costs?” Quest, September-October 2003

Prescription Benefits — A Consumer’s Guide to Free and Discount Prescription Drugs, by Harry P. Thal, Benefits Publications, 2003

County or State Department of Health and Human Services, and local social service agencies

Health Canada

“Guidance Document on Commercial Importation and Exportation of Drugs in Dosage Form Under the Food and Drugs Act”

hc-sc.gc.ca/dhp-mps/compli-conform/import-export/gui-0084_biu-uif-eng.php

Medicare Prescription Drug Plan
healthdecisions.org/guide

Medicine Program
(800) 921-0072
themedicineprogram.com

National Association of Counties
(877) 321-2652
naco.advancerx.com
The website lists counties that have prescription assistance programs.

National Center for Complementary and Alternative Medicine
(888) 644-6226
nccam.nih.gov

National Council on Aging
BenefitsCheckUp
benefitscheckuprx.org
Provides information about prescription programs for people over 55.

Partnership for Prescription Assistance
(888) 477-2669
pparx.org

The partnership is a clearinghouse of more than 475 public and private prescription assistance programs.

Medicare Rx-Connect
maprx.info
Information about Medicare Part D.

Volunteers in Health Care
“Are You Looking for Affordable Medications?”
“Comparative Chart of Pharmaceutical Manufacturers Drug Discount Cards”
(800) 222-6885
rxassist.org

Low-cost sources for equipment

“Use It and Pass It Along,” Quest January-February 2008

“Keep On Keepin’ On: Wheelchair Control Upgrades Keep You Moving,” MDA/ALS News magazine, January 2008

“When Buying Used, How to Be a Savvy Shopper,” Quest, November-December 2007

“Paying for Assistive Tech,” MDA/ALS News magazine, January 2007

“Used, But Not Used Up.” Quest, June 1999

Secondhand equipment:
craigslist.org
disability-resource.com
freecycle.org

RESNA — Alternative Financing Technical Assistance Project
(703) 524-6686
resnaprojects.org/afp

State Departments of Vocational Rehabilitation
jan.wvu.edu/sbses/vocrehab.htm

USA Tech Guide
usatechguide.org

U.S. Department of Veterans Affairs
va.gov/health

Vehicles

See these articles for information on extended-term financing, tech loans, rentals and reimbursement programs.

“Accessible Vehicles Q&A,” Quest, July-September 2009

“In the Market for New Wheels? 10 Tips for Acquiring an Adapted Vehicle,” MDA/ALS News magazine, February 2008

“Programs Make Vehicles Affordable,” MDA/ALS News magazine, June-July 2006

“Show Me the Money: Financing Options Can Make Van Buyers Good to Go,” Quest, June-July 2006

National Mobility Equipment Dealers Association
(800) 833-0427
nmeda.com

Disabled Dealer Magazine
disableddealer.com

RESNA Alternative Financing Technical Assistance Project
(703) 524-6686
resnaprojects.org/afp

Organizing financial information and making decisions

Medical and legal records

Keep complete, detailed records of all medical events in the life of the person with ALS, including appointments, medications, treatments, tests, equipment and hospitalizations. This personal health record also should include names and contact information of the ALS clinic coordinator, nurse and doctor, insurance ID numbers, blood type, a list of medications and equipment, whom to call in an emergency, special care issues and other relevant documents. This will be helpful in speaking with new medical personnel, hired caregivers, hospital staff and insurance representatives. Bring this record along when your loved one has to go to the emergency room.

It's also good to have all important legal papers related to your loved one's care in one place for easy access. A notebook or box could contain: insurance policies, living will, DNR orders, will, names and contact information for doctors, property deeds and other legal documents. In addition, it could include a list of people to be contacted at various times in the disease, where to find other important papers and anything else related to final wishes (see Chapter 9).

Put original legal documents in a safe deposit box. Keep a set of copies in a handy file to take to the hospital or emergency room, and another to take in case of emergency or natural disaster (see "Emergency preparation," page 39).

Planning

ALS brings to the foreground many issues that most of us put off dealing with. Sometime early in the disease, the person with ALS and the family caregiver should begin to plan for later stages of ALS and issues that will be important after death. It's helpful to start early sorting out the financial and insurance questions covered in this chapter. Although absolute decisions don't have to be made about late-stage care, a discussion should begin; it will become harder to talk about when your loved one's speech becomes affected by ALS.

One couple sent their children out for the evening with money for pizza and a movie. Together the two went over all the financial and legal decisions and information

“I organized my papers with a small group of trusted friends. It sounds like a lot of stuff to put together but we did it, had our little meeting to talk things out. Everyone felt good after that meeting that they know exactly what to do whenever something might happen. I feel more at ease, too, that I know everything has been made easy for those I leave to deal with things when I can't do them for myself. **”**

they needed to deal with. They grieved and cried at the thoughts these discussions brought up, but afterwards, having the paperwork done and the ordeal behind them allowed them to focus on day-to-day caregiving issues with greater confidence and calmness.

One early decision is when to transfer titles for property into the spouse's or other relative's name, rather than continuing joint ownership. This will simplify applying for government benefits, collecting insurance benefits and dealing with probate. Be sure safe deposit boxes can be accessed by the surviving caregiver alone.

Always consult an attorney or financial adviser to be sure you understand all the choices and implications; it will cost more upfront but can save thousands of dollars in the long run.

Guardianship of children

Custody of minor children is usually awarded to the other parent or nearest relative. A single parent with ALS who wants someone else to take custody should get the legal documentation completed.

It's also wise to set up trusts or life insurance for later support of the children. Look into **special needs trusts** if a child has a disability.

Power of attorney

A **durable power of attorney for health care** confers on a close relative or friend the legal power to make medical decisions in the event the person with ALS becomes unable to communicate wishes. A **general power of attorney** can grant someone the authority to make legal, financial

Health Care Directive for the Individual with Amyotrophic Lateral Sclerosis

by Lawrence H. Phillips II, M.D.

Recognizing that respiratory failure and/or inability to take in adequate nutrition is frequently the cause of death in cases of ALS, I hereby wish to state in advance my preference regarding invasive mechanical ventilation and feeding gastrostomy tube placement. It is my desire that these preferences guide the decision making of my family and my physician(s) in the event that I am unable to participate in a meaningful way in discussions regarding my health care. I understand that none of the choices made here will be put into effect without my agreement as long as I retain the capacity for decision making and the ability to communicate, in some form, those decisions.

I. With regard to invasive mechanical ventilation requiring endotracheal intubation, it is my preference that:

(Choose one of the following three main options):

A. Invasive mechanical ventilation not be instituted under any circumstances. I understand that such a choice will almost certainly mean that my death will occur earlier than if such support is instituted. I also understand that some processes that might precipitate respiratory failure may be readily reversible and that, therefore, mechanical ventilation may not necessarily be long-term, yet I still do not wish to undergo mechanical ventilation even in such circumstances.

B. Invasive mechanical ventilation be used only when, in the judgment of appropriate medical personnel, the acute cause of respiratory failure is believed to be likely reversible, for example, in the case of choking. If, on the other hand, respiratory failure is a result of the irreversible

deterioration from ALS, I do not wish to undergo mechanical ventilation, knowing that such a choice will almost certainly mean that my death will occur earlier than if such support is instituted.

If invasive mechanical ventilation is used and it becomes evident that long-term mechanical ventilation is required, then (choose none, one, or more of the following):

1. I wish for mechanical ventilation to be discontinued regardless of the circumstances, knowing that this will result in my death.
2. I wish for mechanical ventilation to be discontinued if I should be diagnosed in writing by two physicians to be in a permanent unconscious condition.
3. I wish for mechanical ventilation to be discontinued if I become permanently unable to effectively communicate ("locked-in").
4. I wish for mechanical ventilation to be discontinued if I am unable to return to living at home.
5. I wish for mechanical ventilation to be discontinued if my care results in major financial hardship or other burden on my family.

C. Invasive mechanical ventilation should be instituted in all circumstances for respiratory failure not treatable by other measures, and long-term mechanical ventilation with tracheostomy should be continued with the following exceptions (choose none, one, or more of the following):

1. I wish for mechanical ventilation to be discontinued if I should be diagnosed in writing by two physicians to be in a permanent unconscious condition.
2. I wish for mechanical ventilation to be discontinued if I become permanently unable to effectively communicate ("locked-in").
3. I wish for mechanical ventilation to be discontinued if I am unable to return to living at home.
4. I wish for mechanical ventilation to be discontinued if my care results in major financial hardship or other burden on my family.

II. With regard to nutrition provided by feeding percutaneous gastrostomy tube (PEG), it is my preference that:
(Choose one of the following two main options):

- A. I do not wish placement of a feeding gastrostomy tube at any time during the course of my illness.
- B. I wish for placement of a feeding gastrostomy tube at a time when it is necessary to provide me with nutrition and medications, as determined by my physician, regardless of my choice concerning invasive ventilation. It should be continued with the following exceptions (choose none, one, or more of the following):
1. I wish for gastrostomy tube feeding to be discontinued regardless of the circumstances, knowing that this will result in my death.
 2. I wish for gastrostomy tube feeding to be discontinued if I should be diagnosed in writing by two physicians to be in a permanent unconscious condition.
 3. I wish for gastrostomy tube feeding to be discontinued if I become permanently unable to effectively communicate ("locked in").
 4. I wish for gastrostomy tube feeding to be discontinued if I am unable to return to living at home.
 5. I wish for gastrostomy tube feeding to be discontinued if my care results in major financial hardship or other burden on my family.

In all cases where I choose not to start or to discontinue mechanical ventilation or nutrition via gastrostomy tube, I instruct my physician to provide me with adequate medication to relieve anxiety and discomfort that may occur during the final course of my disease.

(Used with permission of the Journal of Clinical Neuromuscular Disease. 3(3):116-121, March 2002)

“ It’s important to have an attorney who specializes in Medicare, Medicaid and your state programs review your decisions. They can help you avoid pitfalls. It’s important to have second and third choices for power of attorney, executors, and guardians for children in case your first choice can’t do the job when the time comes. Your main caregiver may be too burned out after your passing to take on being executor, for example. ”

“ I can’t stress too much how helpful it was that we had our discussions before a crisis and that we listened to one another rather than what a bunch of preprinted attorney forms indicated was best. ”

“ We made a 4x6 card for [my husband] that included his name, date of birth and photo. We wrote wife’s, best friend’s and doctor’s phone numbers. Disability: ALS — ‘Lou Gehrig’s Disease.’ In red we wrote: ‘No chemical or mechanical resuscitation. No intubation or ventilation, nasal canula, O₂ only. Heimlich or cricothyrotomy only for temporary obstruction.’ We then had it signed by his physician, notarized and laminated. There were copies in the van, wheelchair, friend’s wallet, purse, etc. The EMS system was also made aware. ”

“ [My husband] had a living will, giving me medical power of attorney when he could no longer make decisions. He did stipulate in writing how far he wished his care to go, which was notarized and witnessed by our attorney. This also protected me from prying family members who didn’t like his course of treatment. His wishes were clear, concise and followed to the letter. ”

and other decisions for the person with ALS. These documents are available at most hospitals, from attorneys, in public libraries and online.

When people with ALS can no longer sign their names, caregivers can do it for them. For official documents such as tax returns or property deeds, a power of attorney form is needed in order to sign for a loved one.

Advance directives and living wills

As ALS progresses, it will be necessary to discuss your loved one's wishes regarding advanced medical intervention. When ready, make these wishes formal in a living will and a durable power of attorney. The person may wish to stop ventilatory support when quality of life seems too poor to continue. The person still can receive palliative care at home, in the hospital or in a hospice setting, which alleviates symptoms and maintains comfort. As difficult as it is to discuss such things, it's better to have wishes out in the open while the person is able to communicate; be sure to inform the health care team of the decision. See "Stopping ventilatory support," page 73.

A *living will* (or *medical directive* or *health care proxy*) specifies exactly what types of advanced medical intervention a person with ALS does and doesn't want, should a medical emergency arise or should the person become permanently unconscious. It also can state the person's wishes in case of advanced dementia or other specific crises. These may involve decisions about invasive ventilation or resuscitation. Once a living will is signed, it can always be changed, or overridden by the person with ALS. Having it in place eases family members' uncertainties about unwanted medical intrusions.

While all those close to the person may want to express their wishes, the fact is that interventions or lack of them are ultimately up to the person with ALS (see Chapter 9). He or she should make sure that at least one close loved one knows exactly what they want and has power of attorney.

DNRs

Most people feel very strongly that they want to have some control over the manner of their deaths, if possible. A living will can include specific instructions about DNR (do not resuscitate) orders. These should explain what the medi-

cal condition is, and whether the person can speak, can breathe independently, has a feeding tube, and the location of other instructions. DNR details must fit the requirements of the state of residence. Check to see whether a doctor's signature and/or notarization are required.

A living will or DNR should be very specific. It may have to be read in the hospital, or by emergency personnel coming to the home, usually in a time of crisis when everyone is upset. Generally, "being DNR" means not having any heroic, major interventions done; the hospital still will give comfort care which consists of oxygen, morphine and food. Be specific about what "heroic interventions" and comfort care are requested, and which are forbidden.

Among the life-sustaining procedures that may be applied are:

- CPR (chest compressions)
- defibrillation (shocking the heart)
- advanced cardiac life support medications
- artificial food and hydration
- antibiotics to treat an infection
- artificial ventilation
- pain medication

If the living will doesn't state a preference about food or oxygen, the family can speak to the doctors about what they believe their loved one would have wanted. Doctors often will heed family members' wishes. But if there's disagreement among family members, medical staff will side with intervention. That's why the patient's wishes should be stated in a legal DNR.

On the other hand, emergency medical personnel may not be allowed to honor a "verbal DNR" from a family member. Valid, signed documentation should be produced when the paramedics arrive. In case of confusion, the patient can have a pendant or Medic Alert bracelet attached to clothing that states DNR terms or asks for CPR (be sure the state considers these valid). Some EMTs are trained to look on the refrigerator for a packet containing health history, meds and advance directives, or a notice of where to find these things. If instructions aren't clear and immediately visible, EMTs will do whatever they can to save the patient.

As long as the person with ALS can express wishes verbally, he or she can request that machines be turned off. But usually when this crisis stage is reached, the person is unable to speak or write. A living will can state under what circumstances the person wants lifesaving equipment turned off.

Wills and estate planning

1. Make sure the wills of both the person with ALS and the spouse are up to date and accurately reflect their wishes.
2. Review life insurance policies. Make sure the right beneficiaries are named.
3. Set up trusts, life insurance or special needs trusts to support surviving children.
4. If the person wishes to leave a gift to a charity, under some circumstances *charitable gift annuities* can provide income for the family and leave the remainder to the charity. (To find out about MDA's charitable gift annuities, call the planned giving department at 800-223-6011 or email philanthropy@mdausa.org.)
5. Review all documents periodically with the person with ALS and make any requested changes.

Resources

Organizing financial information and making decisions

Medical and legal records

National Association of Professional Organizers
(856) 380-6828
napo.net

Personal Records Organizer
proorganizer.com

Planning — legal help

Disability at the Dawn of the 21st Century and the State of the States, ed. by David Braddock, AAMR, 2002

Legal Rights of the Catastrophically Ill and Injured: A Family Guide, by Joseph L. Romano, Esq., Attorney, 1998

American Bar Association
apps.americanbar.org/legalservices/findlegalhelp/home.cfm
Consumer guide to finding free legal help (by state). Also check with county bar association.

Elder Law Answers
(866) 267-0947
elderlawanswers.com

LawHelp
<http://lawhelp.org>

National Academy of Elder Law Attorneys
(520) 881-4005
naela.org

Advance directives, living wills and power of attorney

"Five Wishes to Make in Advance," MDA/ALS News-magazine, June 2005

"Stay in Control by Making Advance Directives," MDA/ALS News-magazine, April 2004

"Plan Ahead for Disability with a Power of Attorney," MDA/ALS News-magazine, March 2004

Palliative Care in ALS: From Diagnosis to Bereavement, ed. by David Oliver, Gian Domenico Borasio and Declan Walsh, Oxford University Press, 2006

Who's Right? Whose Right?, Seeking Answers and Dignity in the Debate over the Right to Die, ed. by Robert C. Horn III, DC Press, 2001

Aging with Dignity
"Five Wishes Living Will" document
(888) 594-7437
agingwithdignity.org

Caring Connections
Helpline (800) 658-8898
caringinfo.org

Advance care planning, caregiving, hospice and palliative care, pain, grief and loss, and financial issues. Advanced Directives for all states.

Get Palliative Care

(212) 201-2675

getpalliativecare.org

Includes a palliative care provider directory, a detailed description of palliative care, direct links to palliative care-related organizations and more.

Also see “Hospice” resources, page 143 and 171.

DNRs

At the Close of Day: A Person-Centered Guidebook on End-of-Life Care, by Lance Davis, M.D., and Albert Keller, D.Min., Streamline, 2005
atthecloseofday.com

Caring Connections

caringinfo.org/PlanningAhead

Family Doctor

familydoctor.org

Chapter 8

Finding Caregiving Help



Ask and Receive

One theme is sounded repeatedly by caregivers of loved ones with ALS: *You can't do it alone. Get help!*

Physical demands, emotional demands, other family responsibilities, jobs, and more, eventually take a toll on even the strongest, most devoted caregivers. Don't wait until stress becomes extreme. Take the advice of experienced caregivers: *Get help early, get it from every possible source, accept all offers!* This chapter presents some of the many types of help an ALS caregiver may find valuable.

Some of the help you'll need will be financial (see Chapter 7); some will be assistance with caregiving chores; some will be in the form of medical and other expert advice. Emotional support and respite for the caregiver are absolutely essential forms of help.

Rarely will someone provide the kind of care that the primary family caregiver can give a loved one, but many people can do some of the chores. To clarify the roles, think of yourself as the *carer*, the one who emotionally cares for (loves) the person who needs you, and the *primary caregiver*. Others who are hired or recruited as volunteers are caregivers or helpers, filling assigned tasks.



“ Unless you take time, attention and determination to refill your cup, you will not be able to give your loved one a drink from your cup. Unless you keep your cup at least partially full, you will be unable to offer much of anything to anyone. You are well worth the effort. ”

“ The trick to finding answers is simple: Ask. People who accept the view that there are no answers will find no answers. ”

Hiring caregivers

Agency help

Home health care agencies provide trained people to assist with daily care in the home. The number of hours these aides work depends on the family's budget, insurance coverage and preferences — it can range from around-the-clock shifts to a few hours a week.

Home health agencies usually are Medicare-certified, meaning they meet federal minimum requirements for patient care and management and therefore can provide Medicare- and Medicaid-covered home health services. These agencies provide skilled services from nurses and closely control what their employees may and may not do, meaning that in some instances they only may be allowed to provide medical-related services for the patient.

Staffing and private-duty agencies generally provide nursing services. Most states don't require these agencies to be licensed or meet regulatory requirements.

Homemaker and home care aide agencies offer less skilled care, such as meal preparation, bathing, dressing and housekeeping. Personnel are assigned according to the needs and wishes of each client. Some states require these agencies to be licensed and meet minimum standards established by the state.

Registries or visiting nurse associations serve as employment agencies for home care nurses and aides, matching these providers with clients and collecting

“Once or twice a week I would have my caregiver come from noon until 9 p.m., when [my husband] was put to bed, so that I could get out and have some ‘me’ time. I would take my kids and do something special with them, or even just go to a movie and chill out. Selfish? Sure, but no one else was taking care of me so I had to do it. It also gave me morning time with [my husband] without a caregiver in the house on those days, when we would move at a much slower pace. **”**

finder's fees. These organizations usually aren't licensed or government regulated.

Home care providers are listed in the Yellow Pages under "home care," "hospice" or "nurses." An Area Agency on Aging (see page 169) or United Way chapter also may have a list.

Insurance policies and state Medicaid programs vary in the type of care they'll cover, so be sure to understand the policy's provisions. Some in-home assistance for low-income people over age 60 with disabilities may be covered under the Older Americans Act, with funds administered by the state. A few states allow hiring and paying a family member as a caregiver. Check with your Area Agency on Aging or state department of social services.

Pros and cons of agency help

Nurses and home care aides are trained and experienced in the various chores required to care for a person who's seriously ill. The agency handles all hiring, firing, supervising, payment, taxes and paperwork, and is responsible for finding a person the family likes.

But in-home care is expensive. Users pay a fee that covers the employee's wages plus agency expenses and profit. At the national average of \$18 an hour, this can add up to \$144 for an eight-hour day, or nearly \$13,000 a month for round-the-clock care. The degree of insurance coverage may be a deciding factor in whether to use agency help and for how many hours.

A hired caregiver should behave as if he or she is working for the family, not the agency — willing to learn about

the family's preferences, and respectful of physical and emotional boundaries. The aide should be businesslike and competent but compassionate, willing to communicate with the person with ALS no matter how difficult, and aware of family members' personal space and emotions. If the aide is at all uncomfortable with performing any of the duties required, especially duties related to the personal care of your loved one, don't be afraid to request another aide. Your responsibility is to make sure the aide's behavior gives your loved one a sense that his or her needs will be taken care of competently. This helps reduce the stress of having an outside caregiver in the home.

Occasionally, an agency may send a person who is insensitive to the family's privacy, or indifferent to the patient's personality, dignity and wishes. A paid caregiver even may make crude remarks about death or the person's needs. Or there simply may be a conflict of personality or lifestyle between the worker and the family.

Clarify requirements or preferences with the agency and employee at the beginning of the working relationship. Look at the first few weeks as a trial period; if someone doesn't show up on time, isn't a good fit or behaves offensively, call the agency and ask for another aide. A professional agency will comply without objection.

See "Hospice," page 165, for more about getting care help at home.

Tips for using agency help

- **Be completely honest about needs.** Agencies are familiar with almost every type of assistance required; embarrassment or concern that you're asking too much shouldn't interfere.
- **State preferences from the start.** Be specific about hours, daily routines, and household rules such as "no smoking" and off-limits areas of the home. Don't assume the agency will know your exact wishes.
- **Make it clear who in the family is the authority or decision maker** — the person with ALS or the primary caregiver or both. If everyone in the family gives orders to the employee, the result can be confusion and failure to give proper care. Discuss disagreements among family members outside the employee's presence.

“I'm 24/7/52 for my wife. Cooking the meals, shopping, doing the laundry. I can't leave the house for five minutes without getting someone in. And it can't just be a friendly neighbor — it has to be someone who has done the (ventilator) training course.**”**

— “No Regrets for a Life Less Ordinary,”
Dominion Post, Wellington, New Zealand,
Sept. 1, 2005

“The nurse was extremely nice. She would come out and talk to my parents, just general conversation. She was only there to perform routine blood tests, take vital signs and such, but she spent much more time being a friend.**”**

- **Give feedback to the employee or agency on a timely basis.** If there's a problem, no matter how small, address it immediately to be sure the employee is clear on your wishes. If that doesn't work, call the agency — it's responsible for supervision.

Hiring aides directly

Some families prefer to bypass the agencies and hire a person on their own to assist with caregiving. Someone with little or no medical training can be hired for a lower rate, and can be trained in basic caregiving tasks such as lifting, bathing, maintaining equipment — essentially everything the primary caregiver has been doing.

This type of aide can be a live-in assistant or someone who comes in a couple of afternoons a week to relieve the primary caregiver.

Pros and cons of hiring directly:

- **Pro:** Caregivers can pay a lower rate to someone who doesn't come through an agency, especially a part-time high school or college student.
- **Pro:** There's no need to deal with agency guidelines, regulations and applications.

- **Con:** The person won't have been evaluated by an agency, making the primary caregiver responsible for checking out applicants' backgrounds and ensuring the person is honest and qualified.
- **Con:** Some recordkeeping is involved, primarily hours and payment history. You also may be responsible for paying federal and state employment taxes if payment to the aide is over a certain dollar threshold.
- **Con:** Health insurance may not cover the cost of an independently hired aide (although it may not cover agency help either).
- **Con:** If there's only one helper, there's no agency to send a backup person when the helper is unavailable.
- **Con:** If the helper is a family member or friend, there can be awkward consequences if things don't work out or problems arise.

Finding an aide

There are many ways to find potential assistants. Word-of-mouth is the best resource; ask your MDA support group, faith community, friends and neighbors. Other ways to locate applicants are:

- Check with senior centers or independent living centers.
- Place classified ads in local or college newspapers. College students, high schoolers or Scouts may be able to fill a community service requirement by caregiving.
- Put announcements on bulletin boards or newsletters at hospitals or social service organizations.
- Ask at community college or university career centers or departments of nursing, physical therapy or social work. Students may be looking for part-time jobs or unpaid internships that offer experience.

“When hiring outside help I ask for kindness, attentiveness, gentleness and respect for [my husband] and his changing needs. I need people who have the hearts and inclination to get to know this wonderful man.**”**

- Ask the social worker or other health care professionals at your MDA clinic for other resources in your area.
- Your children's teachers or school administrators may know of parents looking for work.
- Call the community volunteer center.
- Ask at privately owned drugstores and medical clinics.

Deciding what help you need

Before you look for an aide, know exactly what you need the person to do. Make a list of tasks and write a job description. Take a few days to jot down any chores that you could use help with as you go through your routine. Are you primarily interested in physical care, companionship, housekeeping or a combination of all three?

In the job description, include rules such as no smoking, no cooking, no loud music, etc. Do you need the person to have a car to do errands, or can they use yours? Do you need someone to be on call or to work set hours?

Review these guidelines with the person with ALS. Is a male or female assistant preferred? What other preferences does the person have? The loved one may be resistant to having "strangers" come in, but go ahead and look for helpers anyway. It may take some time to convince your loved one to allow others to provide personal care, but at a certain point the primary caregiver knows best what's needed in the house.

Interviewing and hiring

Hiring your own caregiving assistant means you have to screen applicants for criminal records, training, job referrals, etc. You can't take a chance on allowing someone dangerous, incompetent or dishonest into your home to deal with a vulnerable person.

When someone calls in response to the ad or inquiry, ask what they'd like to know. For example, it's a bad sign if their first question is "How much does the job pay?" or "How much time off do I get?" It's better if they ask what some of the duties are or how many hours they'd be needed.

Explain the job briefly, but don't describe your loved one's exact physical condition or the number of people in the

household. There are criminals who search out vulnerable victims from ads and notices.

You can ask a bit about experience or background over the phone; the phone call should let you weed out inappropriate applicants. If you get a good feeling from the phone call, ask the person to come for an interview. You may want to interview three to five people before choosing one.

Expect interviewees to dress appropriately (that doesn't mean hose and heels, but they should at least be clean, neat and modest) and to arrive on time or call if they must be late. Expect someone who listens, makes good eye contact and asks relevant questions. Most applicants should bring a résumé, though a young student or a person just re-entering the job market may not have one.

You can print out a simple job application form from the Internet or create one.

Ask some open-ended questions: *Tell me about yourself. Why are you interested in this job? What's your school/work schedule? What do you do in your spare time?* The answers will give some insight into communication skills and personality, important factors in a future relationship.

Of course, ask about experience — even volunteer or family caregiving experience could be valuable. If they've done in-home care before, ask why the job ended.

Explain your needs in some detail and find out if they have experience bathing, feeding, etc. Describe a typical day and what's expected of them, and check the reaction. Be sure to tell them of any uncomfortable tasks you expect of them, and ask if they have the physical strength for lifting and transferring.

If you're seeking a live-in, be specific about guidelines: Can anyone move in with them? Can they bring a pet? What's included — rent, utilities, food, phone, cable, furniture? Are days off set or negotiable? Can they have friends over for dinner or overnight? Ask if the applicant is in a relationship, and make clear whether it's OK for the partner to spend time at your home. If so, how often?

For all applicants, ask for names and contact information of several people you can call for references. Former employers are best; students also may list teachers or people from church or the neighborhood for character references.

If at any point in the interview, you know the person is unacceptable, just conclude the meeting. There's no need to waste time talking with someone you aren't going to hire.

When you've narrowed the field to one or two leading candidates, it's time to check background. This is imperative, especially if the person hasn't been referred by someone you know. You can't be too careful about exposing your home and your loved one to someone you don't really know. Make sure they give you a permanent address, date of birth, Social Security number and driver's license number.

Run a criminal background check and driving records check. There are several websites that will do this for a fee or will explain ways to go about searching; see lexisnexis.com; virtualchase.com; searchsystems.net. Some search programs are little better than scams. Among the good ones are NetDetective.net, ReverseRecords.org and CourtRecords.org.

Introduce the person with ALS and get his or her reactions. Trust your instincts.

Managing caregivers and helpers

- Inviting someone to spend many hours in your home isn't easy. First, all those involved must accept that the assistance is needed enough to sacrifice a bit of privacy. Eventually everyone, including the person with ALS and the primary caregiver, will come to accept that this helper can improve the family's quality of life.
- To make things as easy as possible for everyone, give the aide as much information as possible. Offer a written summary about ALS and your loved one's condition, and provide MDA materials on the disease.
- Post a daily schedule, listing times for getting up, bathing, meals, meds, etc. Be very specific about dosages of medications. A monthly or weekly calendar that lists times of therapy and doctor appointments also would help. If the person with ALS has speech difficulties, these lists will help new caregivers understand the needs he or she may be describing. Explain any other keys to assist with communication.
- Make copies of any instructions or advice you've received from health care professionals, such as how

to handle the ventilator. Go through each new chore carefully; demonstrate several times and show the new person how to watch for your loved one's reactions. Watch as the aide performs the task the first few times, and give helpful hints. Be patient with the learning curve.

- Ask the person with ALS for feedback and make it clear to the aide.
- If your loved one is reluctant to be assisted by others, explain that accepting someone else's help is a great way for your loved one to help you. Their resistance may be a form of denial of the seriousness of ALS; requiring outside help means admitting the depth of the problem.
- Let the helper take on chores gradually or assist you until the person with ALS gets used to the idea. Explain to your loved one what you will be doing while you're gone, and make it clear you'll be back. Remind the aide to be patient.
- If you aren't going through an agency, you may wish to draw up an informal contract with the aide that spells out hours, pay rates and other details of the job. You each should sign two copies, and each keep a signed copy.
- For aides you hire directly, check with an accountant to be sure you're keeping the correct records. Record hours and payments. Tell aides that, as contractors, they're responsible for paying their own Social Security taxes. Some people have the aide sign a time sheet or pay sheet every week or two weeks, and give them a copy of the record to keep.
- At the end of the year, send a W-2 form to the IRS indicating how much you've paid for income tax purposes; the worker also gets a copy.
- Your homeowner's insurance should cover liability for injury. If you're concerned, ask the person to sign a no-fault agreement.

“Perhaps you can explain to him that, although physically caring for him is a way to be together and gives you both satisfaction (and avoids having strangers in the house), it is very tiring for you and will become extremely difficult in the (hopefully) long run. Also, it will leave less time and energy for the more pleasant things you usually do together. **”**

“My mom has had ALS for four years and is opposed to much evening help from outside the family. Now, as things get worse I fear my dad and I are not enough help throughout the evenings and weekends. I spend around 30 hours a week with her now, outside of work and my own family, and am getting worn out. How do I make her see we need help, but that I am not abandoning her? **”**

“My mother does not refuse help because she knows how much we need it, but she doesn't like having someone around all day. We hired a caregiver that is around the same age as my mother and has similar interests. When my mom wants to be alone she just lets her know, and the lady will go in the other room, read a book or watch TV. **”**

Asking for help from friends and family

Most of us find it difficult to ask for help; we like to show the world, and ourselves, that we can handle it, whatever it may be. ALS caregiving is such a personal, round-the-clock experience, it's difficult to know how to parcel out chores.

There are many ways others can help. Friends, neighbors, church members, co-workers, social acquaintances and family members may have offered to help, but they aren't going to line up at the door. They're at home waiting for a call: People usually will help if they receive specific direc-

tions. Be aware of your own psychological, physical and mental strengths, and look for friends to help in the areas where you're less confident.

Using volunteer help from those who care about your family has many advantages:

- It saves money.
- It draws friends and family closer together and makes everyone feel needed (see “Rewards of Caregiving,” page 181).
- It gives the primary caregiver a little respite, which will benefit his or her physical and mental health.
- Knowing there's help can make it easier to keep a loved one at home, avoiding or delaying placement in a care facility.

List all the people who have offered to help, and all those who haven't offered but who might be willing if asked. Develop a chore list, including frequency, and start matching chores with available people.

For a more formal way of organizing volunteer helpers, see “Share the Care circles,” page 163.

Friends and neighbors don't need to get involved in the most intimate aspects of caregiving. Household chores, errands, child care and such are more appropriate and comfortable for them, and they'll allow the primary caregiver more time for hands-on, one-to-one care. In requesting that others rake the leaves, drive the kids to soccer, pick up groceries, etc., you'll have to let go of total control over these tasks. You don't have the energy to sweat the details, and your way isn't the only way. If it gets done, you don't have to worry about it anymore.

Here are some other tips for getting friends, family members and neighbors to help:

- **Be clear about what you want.** Avoid vague requests: “Maybe sometime you could stop by to visit with Al for a few minutes?” Instead be direct and specific: “Could you come by Sunday and watch the game with Al on our TV, so I can take a nap upstairs? And please bring some snacks.”
- **Brainstorm sources of help and expertise.** Be careful not to ask the same people over and over. Think about neighbors and acquaintances who might be glad to

do simple repairs or stop by the dry cleaners. Ask a knowledgeable relative to help sort out bills or insurance papers.

- **Get help getting help.** Widen your list of helpers by asking a close friend or relative to ask people on your behalf. Using an intermediary often makes the process more comfortable for everyone.
- **Set up a website** such as MDA's *myMuscleTeam* care coordination site (mda.org/mymuscleteam), which provides an efficient, effective way to recruit and schedule volunteer help from family and friends.
- **Keep lists of chores by the phone or in a purse**, so when well-wishers ask, "Is there anything I can do to help?" you can tell them. Or, ask if you can call them when you need something from the store or a short respite. If they say yes, call them.
- **Expect some hesitation or refusal.** Your request simply may come at an inconvenient time. Consider giving them a second chance, but don't spend time trying to convince someone who's refused two or three times, or doesn't keep a promise.
- **Accept gifts.** Some people may not have time to offer but would be glad to spend some money. If asked, sug-



“In a rural town there are few services available. In order to manage your daily life with ALS, you need to use all available resources. When needed, family and community members run errands and take our two teenagers to high school when it rains and the school bus won't tackle the long and bumpy dirt road.

If you don't have an extensive support system in place, already (i.e., friends, family, a close-knit community), then either move or start making friends. This is not a disease that can be battled alone.”

“We were lucky enough to have two dear friends who not only brought meals twice a week, but they stayed to keep us company while we ate, feeding [my husband] so I could enjoy a hot meal, then cleaned my kitchen for me while I put him to bed. Then they would make a pot of coffee and sit and visit with me for a few minutes once things quieted down. They never stayed more than two hours and seemed to know which days the meals were most needed.”

“Tell people what you need when they offer help. Even if it's a gallon of milk, let them get it for you. One less thing to fret over.”

“If you give people a task they're suited for and are comfortable doing, they lose their fear of getting involved. Give them a group to work within, a structure to fit into. Give them an opening to say yes or no — and oftentimes they come back with an even better suggestion of a way they can help.”

gest a book of postage stamps, an in-home massage, a takeout meal, a professional housecleaning, or a movie or book on tape or disk.

- **Don't get bossy.** If you find you're ordering your friends around and expecting them to do things in certain ways or take on additional chores, stop and think. Either let the person do the task in his or her own way, or realize

that if you need that much control, it would be better to hire someone who expects to take orders.

Children as helpers

Even very young children like to feel they're helping their loved one with ALS. Children have helped parents with ALS with suctioning, feeding tubes, changing trach tubes

How to Cope: Just Say Yes

by Alan Coleman

From the beginning, remember this: Yes is not a dirty word. There is one question that you will hear often, "Is there anything I can do to help?" You need to get in the habit of answering that question with Yes. There are two reasons to do so.

1. At the beginning of your caregiving experience, it might be that you don't actually need the person's help. In fact, you may feel that accepting their offer will impose upon them — even though they volunteered the offer. But the people that love you and your loved one will feel an overwhelming sense of helplessness regarding your loved one's condition and they truly want to help. They actually need to help you more than you need their help, so start saying Yes from the beginning. Don't deny them the privilege and satisfaction of taking part in your caregiving responsibilities.

2. Even if you do not need the help initially, your loved one's condition may eventually reach a point that extra help will be necessary to care for them properly. If you're already in the habit of saying Yes, then it will be natural to continue doing so at that point of need. If you haven't been accepting offers of help, it can be difficult to start ... especially with people that you may have declined several times. In fact, if turned down a couple of times, some people will quit offering because they don't want to seem intrusive.

Just say Yes ... it'll help everybody involved.

Make a list

Not only do you need to say Yes, but you need to be able to answer the next question you'll hear: "Okay, what

can I do?" To answer this question, you need to make a list. Before you start your list, understand this: the help you get from others DOES NOT have to be direct care for your loved one. It can be anything you do in your daily routine that can be done by someone else. When you start your list, think of it as a diary of everything you do for a week ... and I do mean EVERYTHING. Start in the morning when you first get out of bed and list everything you do for your loved one AND everything you do for yourself all day long until you're back in bed at night. As the week goes on, you can leave off things that you've already put on the list. For instance, if you go to the mailbox every day, just list it one time. At the end of the week, eliminate any items that repeat.

Then you are going to remove anything on the list that is physically impossible for someone else to do. For instance, nobody but you can go to the bathroom for you (remember, I said EVERYTHING goes on the list). Next, remove anything from the list that your loved one simply prefers that you do. There were some bathroom chores that my wife, Dink, didn't mind other people doing for her and those stayed on the list. There were other bathroom chores that Dink did not feel comfortable with anyone BUT me doing, so those things came off the list.

What you should have at this point is a list ... preferably a long one ... of everything you do that can be done by someone else. Actually, what you have is the initial list. You should never consider your list as being "completed." Add to the list constantly as your daily routine changes and always keep a few copies of the list with you so you can give them to people who ask.

Now you're set. When anyone asks what they can do to help, whip out the list and let them choose.

and catheters, or household chores. They can learn by watching and be given simple tasks — “bring Mommy the formula for Daddy’s dinner” — at first.

Children involved in caregiving learn compassion, cooperation, patience and responsibility. Sometimes, however, teens who take primary responsibility for caregiving develop depression, anxiety, insomnia, loss of interest in

Bring in the cavalry

The extent to which you need outside help with your caregiving responsibilities is going to vary depending on the nature of your loved one’s condition. If your loved one has a degenerative disease, like Dink’s ALS, then having outside help will be critical to the success of your caregiving efforts. In fact, you will not be able to do it alone ... there’s just not enough hours in the day and you simply don’t have enough hands! Obviously the solution is to recruit as much help as you can, but what is not so obvious is that you may have to recruit help to handle all the help you get.

We had some wonderful people step to the plate for just that purpose. A lady at our church volunteered to organize all the meals that people brought to us. It was awesome. When anybody came up to me and said they wanted to bring us a meal, I told them to go talk to Judy. I never had to worry about it ... all I had to do was be at home when the meals were delivered. Two other ladies — one from church and one from Dink’s work — volunteered to coordinate the sitters that came in to stay with Dink when she could no longer be alone while I was at work. They handled everything from recruiting to training to scheduling the ladies that we came to call “Dink’s Angels.” There’s no telling how much stress they took off my shoulders as the efforts of all of these people allowed me to concentrate on taking care of Dink.

You can find volunteers among any group of people to which you belong. Your workplace. Your loved one’s workplace. Your church. Social or business clubs. Obviously family — Dink’s entire family gathered monthly to clean our house and do yard work. Just put the word out that you need help, identify key people who will be willing to help you manage the help you get, then turn

school, or delay becoming independent. Teen caregivers need to get in touch with the social worker at the MDA clinic to find help.

Long-distance caregivers

Today, family members are spread across the country, and it isn’t always possible for them to relocate to care

those responsibilities over so you can concentrate on caring for your loved one. Remember, the goal is for your loved one to be cared for, not for you to martyr yourself to their care. Which leads to my next point.

Take care of yourself

This was the hardest thing of all for me to do when I was taking care of Dink. But it’s also the most important thing you can do. If you don’t take care of yourself, you won’t be able to give your loved one the best care you’re capable of. It’s just that simple. I know ... there will be times — many times — that you will feel guilty about doing things that you used to do with your loved one. Or doing things that your loved one simply can’t do anymore. But you have to do those things to keep yourself healthy ... in mind as well as in body.

Arrange times that someone else can stay with your loved one for a few hours while you get away, BUT, don’t use those hours to do things on the list you made! This time is for you to get away, get rested and get rejuvenated. Go for a drive. Go to a park. Go to the library and read something pleasurable. Go for a walk, run, bicycle ride, swim, workout, or whatever else you enjoy doing to keep yourself fit. If you have a hobby, concentrate on it for a while. Go see a sappy movie. What you do is up to you, just be sure that you do something for your own well-being.

I can’t help thinking about those goofy instructions you hear every time you get on an airplane. Put YOUR [oxygen] mask on first, then take care of the other person. It’s important enough to repeat: If you don’t take care of yourself, you can’t take care of someone else.

(Excerpted with permission from Alan Coleman, author of *Firmly in His Hands*.)

for a parent or sibling. Long-distance caregivers have special difficulties:

- They can't see what's really happening or do hands-on care.
- It's difficult to reach doctors or social service agencies and coordinate needed care.
- They may feel guilty and sad over not being involved.
- Traveling back and forth can be exhausting, expensive and frustrating.
- Other relatives, including the one with ALS, may not understand why the person can't just "drop everything" and come to help them.

With good planning and cooperation, even those who are miles away can contribute.

If a long-distance relative is the primary caregiver, a leave of absence from work may be possible. If not, or when young children are in the picture, frequent visits may be the only

“We call often; I maintain a website that I regularly update with photos of the kids, what we're up to, etc. The kids occasionally write emails and send drawings, but I want to do more. We are going to start volunteering; if I can't help out my father-in-law in person, I can at least help a local ALS patient. **”**

“I live in Canada, and my parents are in the States. Although packing up and moving home was all I wanted to do, I knew it wasn't the best thing for my family and two young children. I felt terrible, and struggled with it for so long. I am lucky that as a teacher I can spend a decent amount of time with my parents in the summer. The distance cannot take away from your relationship, and in the end (as ALS teaches us) memories, moments and those important relationships are all that really matter in this world. **”**

way. Try to be there for doctor visits to meet the health care team, or for selection of major equipment.

The caregiving role will require a great deal of communication and coordination with those who are nearer the loved one:

- **Entrust a neighbor or close friend** who lives near the relative to check up and visit on a regular basis. Talk to the person frequently, and be sure they contact you if there are any serious changes.
- **Go to the loved one's town to interview formal services** such as visiting nurses, senior centers, adult day care or a meals program. Keep in regular contact with these agencies and make sure, through a friend or neighbor, that the person with ALS is receiving proper care.
- **Meet the physician** and keep in regular contact. Call and speak to the physician directly, or ask how you can receive regular, updated notes on the visits and tests that are administered.
- **Hire a private care manager** through a social service agency or home health care agency. This person can coordinate services from physicians, nurses, therapists, social workers, homemakers, durable medical equipment and supply dealers, and volunteers. Private care managers can monitor daily care, assist families with implementing a long-term care plan, and keep family members informed if a problem arises.
- **Customize a caregiving network** (see "Share the Care circles," page 163).

Long-distance caregivers also can work with primary caregivers who see the loved one daily.

- **Be informed about ALS** so you understand what's going on.
- **Visit as often as possible** so the family knows they're a priority in your life.
- **Make the moments you are together count.** Think in advance of your visits about things you can do together and ways to help. Just let your loved one enjoy your company.
- **Offer financial support if possible.** If family members are reluctant to take money, offer to pay for a specific need — in-home care, respirator supplies, food, etc.

- **Provide emotional support and involvement.** Phone the loved one often to keep them part of your world, and find out how the day went. Phone the primary caregiver as well and offer support or an ear for their frustrations.
- **Maintain a website with family photos and news, or email pictures often.** Encourage children to send notes and drawings.
- **Respect the primary caregiver's leadership** and offer cooperation; avoid disagreements whenever possible; make gentle, helpful suggestions.
- **Get involved with MDA in your community;** participate in fundraisers; volunteer to help a local family dealing with ALS.

'Share the Care' circles

A unique method of organizing helpers to share the load is known as a Share the Care circle. These are based on the book *Share the Care: How to Organize a Group to Care for Someone Who is Seriously Ill* (see "Resources," page 170).

Share the Care circles can start with a small group, such as a prayer group at church, a handful of co-workers or a bunch of longtime friends. This core group takes on responsibility for organizing needs and finding people to fill them, removing the burden from the primary family caregiver of recruiting and organizing volunteers.

A circle eventually includes dozens of people and reaches far beyond the personal acquaintances of the person with ALS. Long-distance caregivers can play important roles in the circle. Groups often give themselves names: Jeff's

““Being part of the group was the most important caregiving lesson I could ever learn ... how to take care of myself while I took care of others. As I learned to share workloads, decisions, feelings, and responsibilities — to ask for help when I needed it and allow myself to say no sometimes — my life began to return to me.”**”**

— *Share the Care*

by Cappy Capossela and Sheila Warnock,
Simon and Schuster, 2004



Angel Group, David's Circle, Friends of Cindy. Information about needs and schedules can be posted on a website such as MDA's *myMuscleTeam*, so volunteers can offer specific help.

In care circles, a few individuals take responsibility for organizing areas such as laundry, shopping, child care, communication, etc. The circles ripple outward to include all kinds of specialists: attorneys, accountants, fundraisers, building contractors, handymen, equipment donors. As each person is contacted for help, that person reaches out to others. The circle organization assures that no one person has too much responsibility, and everyone has a manageable workload. If the family is so inclined, a Share the Care Circle can be pitched as a human interest story to a local newspaper or TV station. More attention equals more volunteers.

See "Resources" at the end of this chapter for information on how to organize a Share the Care circle; next time someone energetic and committed offers to help, suggest trying this approach.

Other Assistance

Online and delivery help

Many services and businesses offer home delivery and pickup, or online or mail order services: pharmacies, groceries, laundries and dry cleaners, holiday and birthday shoppers, clothing and electronics from department stores, movies and CDs, etc. Ask a good friend to investigate these services and line them up for you. Using these efficiently can save tremendous amounts of time (and

gasoline) that make it worth any extra cost.

The most efficient grocery delivery is to place a large order or a regular weekly order, rather than calling every time you need a gallon of milk. It's possible to order online from local supermarkets or from national services that ship within 24 hours.

Explain your situation to businesses such as car repair shops, etc., and they may be willing to pick up and deliver.

Home modifications

Volunteers can be found to remodel a room, build a ramp, widen doorways or install a roll-in shower. Put the word out among friends and start the search for handymen or contractors who will donate labor and/or materials. Independent living centers (CILs) or home health agencies may know of someone who will help. The Homebuilders Foundation exists in several states as a philanthropic arm of the industry; they may help locate a volunteer contractor.

If it's necessary to move in order to accommodate the person with ALS, or for financial reasons, the state's Social Services or Family Services Department may know of a program to help find housing. Some states are committed to helping people stay in their homes. (Also see Chapter 7 for information about reverse mortgages and low-interest loans for home modification.) CILs also have leads on low-income, accessible housing.

Elder law attorneys or legal aid organizations can help with rental disputes and agreements.

Medical help

An MDA clinic or MDA/ALS center can refer your family to a range of needed assistance. Medical care is coordinated, and clinics are directed and staffed by neuro-muscular disease specialists. The clinic or center can refer you to knowledgeable therapists, social workers, medical specialists and others. The local MDA office can point you to community services such as home health agencies, equipment providers or independent living centers.

The **social worker** at the MDA clinic provides help in navigating the health care system, and is a great source of referrals to family counselors, in-home help agencies and

assisted living facilities. The social worker is the family's advocate or case manager, who can help coordinate the various types of care needed or being received.

However, in the current health care system, families coping with ALS need to be their own advocates. Dealing with medical professionals outside the MDA family, such as pulmonologists, skilled nurses or therapy agencies, can require that families know what's expected and needed, and be prepared to educate the professional.

Understand what each person does in the medical setting so you can ask the right person. Get to know office staff so they'll be more receptive. When asking for information, be firm about what you need and strive to remain calm no

“I have been an R.N. all my adult life, eight years as a hospice nurse, yet never encountered ALS until my dad was diagnosed. Look at the years I didn't have a clue.”

“My sister seems to feel better when she can talk to someone other than family and close friends, such as care workers, the hospice staff and even the chaplain that visits her a few times each week. She can talk openly with people like that, where with family, we always want to hear what WE want to hear, and not always what she wants to say, and that is natural.”

“I tell patients, 'I'm not here to replace your physician. I'm here to augment your physician.' I really encourage patients and family members to call me if they have a concern or they have a worry, if they have a problem. I don't want them sitting on it and not contacting us and just getting anxious when maybe there's a pretty easy solution. If it's a more complex issue, then the patient needs to be seen sooner.”

— Nurse practitioner

matter how frustrating the experience.

Remember that doctors are legally and professionally responsible to your loved one, not to family caregivers, unless a caregiver holds **medical power of attorney** or **durable power of attorney**. Your loved one must communicate to the doctor that the caregiver is to be part of exams, tests, discussions and decision making.

To become informed about providing physical care, ask occupational therapists, physical therapists and others how to do basic things: range-of-motion, transfers, vent maintenance, tube feeding, basic nursing skills, etc. Manufacturers' representatives should be willing to answer questions about equipment, and demonstrations often are given at support groups. Some hospitals teach these skills, as do some home care and hospice services. The American Red Cross has a curriculum on family caregiving skills training.

To learn about medical issues, do online research or even take a brief course. You'll be able to ask the doctor more knowledgeable and pertinent questions and gain a higher level of information.

One woman with ALS wrote up a detailed explanation of the disease, and how it did and didn't affect her. She included her needs and expectations for caregivers. Everyone — aides, physical therapists, nurses, volunteers — who dealt with her was required to read the material. Professionals who thought they knew everything about ALS learned more and got a better understanding of the person's individual needs.

Educating staff about ALS and its effects is especially important when the person is in the hospital and is being treated by nurses, residents and others who may never have helped an ALS patient. A caregiver may have to get "politely pushy." It's better to be considered obnoxious than to let your loved one get the wrong type of care. Ideally, staying in the hospital with your loved one ensures their specific needs are made known and accommodated.

Keep medical information organized and complete so that each new person you see at the clinic, hospital or in your home easily can find answers and data.

Many hospitals have a **patient advocate** who speaks in behalf of patients in order to protect their rights and help

them obtain needed information and services. This often is a nurse, social worker or other health care provider. If there's an ongoing problem with hospital treatment, ask to speak to the patient advocate.

Long-Term Care

At some point, even the most devoted caregiver may need to turn over a significant portion of the loved one's care to professionals. It's emotionally very difficult to place a loved one in a long-term care facility, but his or her medical needs, or a caregiver illness, financial realities or other factors may make this essential.

Most of the time, some percentage of the costs of the facilities described here is covered by private insurance, Medicaid or Medicare, or the person's SSI or SSDI income. Once a person with ALS goes into long-term care, Medicare may no longer provide certain equipment. Check with an expert to learn what equipment should be obtained prior to moving to a long-term care facility.

Hospice

Hospice services can provide a transition from care at home to full-time care away from home. Hospice care is available to people in the end stages of a terminal disease. Hospice staff and volunteers coordinate with the patient,

“The level of care Mom needs now is great and we are drowning a bit trying to work and care for her, too. We called hospice, I asked that they be sensitive as we really haven't discussed all the possible symptoms that she might develop since everyone is so different. **”**

“Get all your equipment before you sign on hospice; insurance won't pay for these items once you're in hospice, but will if you have a home health agency. Some hospices accept people with feeding tubes, BiPAP and CoughAssist but prefer them to be in place. **”**

physicians and family members to provide day-to-day care and comfort, in the home or at other locations. Services include administering medications, including those to ease suffering; performing personal hygiene tasks and minor medical procedures; and offering compassionate companionship and support. Most hospice services provide support to the family as well. The hospice service may provide a lot of equipment for the patient at home, including bed, bedside table, alternating pressure mattress, roll-in shower, commode, medications and other supplies.

In general, hospice patients receive only comfort care and symptom management, not life-prolonging devices or therapies. However, pre-existing feeding tubes are OK and some hospice programs also accept patients who already are vented.

Hospice care often is provided in the home, but can be conducted in hospice centers, hospitals, nursing homes or long-term care facilities. The cost is covered under Medicare, Medicaid and most private insurance plans. (See Chapter 7.)

Call local hospice services when your loved one needs help with breathing, feeding, turning, etc. They can do a home evaluation and explain what services may be available and when. Try to find a hospice whose staff is familiar with ALS.

A 2007 study funded by the National Hospice and Palliative Care Organization found that hospice care seems to prolong the lives of some terminally ill patients by days or even months. Some reasons for this may include:

- More personalized attention may result in improved monitoring and treatment.
- Hospice care offers a comprehensive approach, focusing on the patient's emotional and spiritual needs in addition to physical health.
- Family caregivers are offered support and training. Their reduced stress or workload may help patients feel like less of a burden, and so increase their desire to live.

Assisted living facilities

Assisted living bridges the gap between independent living and nursing homes. Residents in assisted living

centers aren't able to live by themselves but don't require constant care either. This may be a feasible arrangement for someone with ALS for a period of time, though ultimately the progression of the disease will likely require placement in a hospital, hospice or nursing home.

Assisted living facilities offer help with activities of daily living such as eating, bathing, dressing, laundry, housekeeping and taking medications. The facility should create a service plan for each resident, detailing personalized services required by the resident and guaranteed by the facility, and update it regularly. People who live in newer-model assisted living facilities usually have private apartments, and there are common areas for socializing, as well as a central kitchen and dining room for preparing and eating meals.

In 2012, the national average cost of an assisted living facility was \$3,300 per month, which isn't likely to be covered by insurance. Other terms used for assisted living facilities include: residential care, personal care, adult congregate living care, supported care or enhanced care.

Nursing homes

A nursing home or *skilled nursing facility (SNF)*, also sometimes known as a *rest home*, is a place of residence for people who require constant nursing care and have significant deficiencies with activities of daily living. Physical, occupational and other rehabilitative therapies are offered. Nursing homes are designed to care for very frail people who can't care for themselves and have numerous health care requirements.

Inquire about assisted living facilities and nursing homes in the community and be sure they can provide all the care your loved one requires. Be sure they permit ventilators,

“ Ask if a facility has ‘ALS protocols’ — procedures useful in caring for people who are paralyzed, speechless or on a vent, such as Hoyer lift operation or communicating via alphabet board. If not, ask administrators if they’re willing to learn about ALS, make changes and train staff to meet the unique needs of ALS patients. ”



feeding tubes, etc.

After years of caring for a loved one with ALS, having him or her live outside the home is a drastic transition. It's another loss to grieve, and usually isn't a change that anyone welcomes. The caregiver faces the shock of a suddenly changed role, just as the person with ALS may feel abandoned or adrift in unfamiliar surroundings. There's also a sense of relief for the caregiver as duties are reduced, but it comes mixed with sadness, loss and emptiness.

The caregiver and person being cared for can maintain their close relationship after placement in a long-term care facility, and the caregiver will remain vitally involved in care decisions. The loved one still needs a caring person to be sure that things are going well and the right care is being provided. Get to know the staff at the facility to understand who's responsible for what and who is your best ally if there are problems.

One positive aspect is that now time together can be focused on each other, rather than on chores to be performed.

Rewards of Caregiving

Health research scientists are finding that — despite the tremendous burden that caregiving presents — people actually can benefit physically and emotionally from the act of caregiving. (Also see page 25.)

One recent study noted that "individuals who reported providing instrumental support to friends, relatives and neigh-

bors, and individuals who reported providing emotional support to their spouse, had a 30 to 60 percent *decreased* chance of dying over the course of the study," compared to their nonhelping counterparts.

It's believed that the positive emotions generated by helping behavior may bolster caregivers' overall sense of well-being, affecting life satisfaction and the ability to cope with stress. Positive emotions also have been shown to lead to increased longevity and faster recovery from cardiovascular stress.

These effects are available not only to the primary caregivers of people with ALS or those who spend many hours a day caregiving —they also benefit volunteers and others who are asked to help. This "helper's high" can be found in the little moments such as a smile shared between caregiver and loved one. Some may find pride in overcoming difficulties or in knowing a loved one is receiving the best care possible.

Of course, these benefits arise for the caregiver *only* when he or she is given adequate support. So inviting others to share the care not only helps the primary caregiver; it gives others a strong emotional boost — it's a win-win-win situation!

Respite help

One way to experience the rewards of caregiving is to get support through respite care. Call on your network of paid or volunteer helpers to stay with your loved one so you can get a hair cut, go to a ball game, shop, enjoy a hobby, or just sit in the park and watch birds. Ask for babysitting or for someone to take the kids overnight occasionally.

Every community has respite facilities such as adult day care centers that will take the loved one for a few hours or days to give the caregiver a break. In addition, a loved one who qualifies for hospice can spend a night or two at a hospice facility.

Schedule respite regularly — you've earned it, and it will help everyone in the family, including the person with ALS. At first it may feel uncomfortable or "selfish" to take time for yourself, but remember this is necessary for you to do the best job possible over the long haul.

Support groups

MDA support groups are among the most important services the Association provides. Caregivers credit support groups with new friendships, important advice on equipment and more. Couples and individuals who are farther along on the ALS journey give new members a realization that they can get through this sometimes; no professional counseling or family support is quite as meaningful as peer support. People don't have to explain ALS or any of the needed care or devices. They don't have to explain the exhaustion of caregiving or the other emotions and stresses they experience. Everyone who's there already knows.

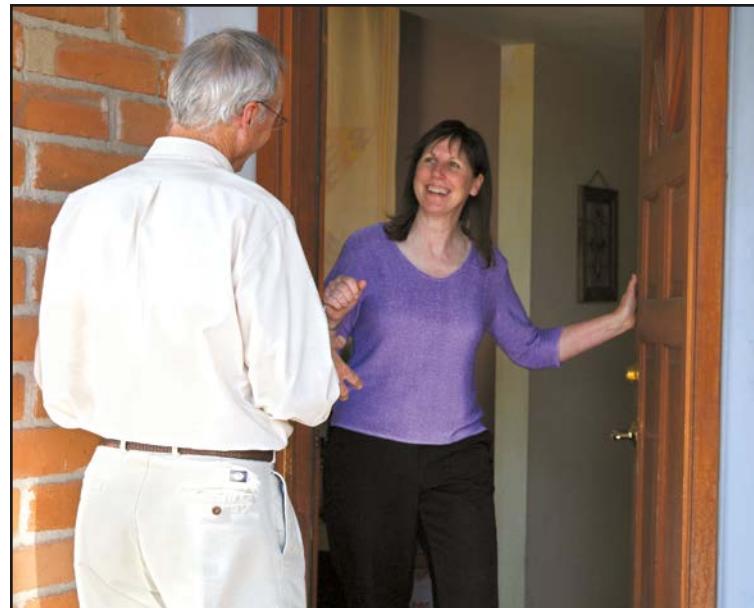
Support groups aren't just places to share emotions and pick each other's brains. Groups often invite knowledgeable speakers or have casual social time. There often are no boundaries on what things can be discussed. Toileting, child-rearing problems, sex, family conflicts, breakdowns, death — all are parts of the ALS experience that can be brought to a support group.

Attending support group meetings can be difficult because of distance or other issues. There are many online listservs where people with ALS and caregivers share viewpoints and questions. See "Resources" for a list. Contact your local MDA office (800-572-1717) to see if they can put you in touch with others in your area who are living with ALS.

Private or family psychological counseling may be helpful for a time and often is covered by insurance. Check under psychologists or social workers in the Yellow Pages, or ask for a referral at the MDA clinic.

“Sometimes life even gets better at a care facility, thanks to activities not available at home, like pool therapy, classes or people to talk to. It’s never easy, but there’s relief in knowing competent care is available when needed.”

“Planning ahead is the best way to land in the best spot. When you know you have ALS, get on waiting lists at many assisted living and nursing homes.”



Resources

Back issues of MDA publications such as Quest and the MDA/ALS Newsmagazine can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Help for caregivers — general

See Chapter 1 Resources, page 27.

“Family and Caregiving Resources for People with ALS,” MDA/ALS Newsmagazine, May 2007

“Searching for Information on ALS and Caregiving?” MDA/ALS Newsmagazine, January 2005

“Family Caregivers: 10 Ways to Get More Help,” MDA/ALS Newsmagazine, November 2004

“Reducing Caregiver Stress May Help Loved One’s Depression,” MDA/ALS Newsmagazine, February 2003

“Caregiver Support Programs Innovative — But Going Nowhere,” MDA/ALS Newsmagazine, January 2003

“Families Take on Responsibility,” MDA/ALS Newsmagazine, January 2003

The Comfort of Home: An Illustrated Step-by-Step Guide for Caregivers, by Maria Meyer and Paula Derr, Caretrust Publications, 2002

American Medical Association Guide to Home Caregiving,
by the American Medical Association, Wiley, 2001

CareCircle
(914) 741-6742
carecircle.com

MetLife Mature Market Institute
metlife.com/mmi
This site offers many resources for caregivers.

Local community services

Check county and state government listings for:

- Health and Human Services Department
- Public Health Department
- Social Security Administration
- Mental Health Department

Contact the social service departments of hospitals and clinics. Locate adult day care centers and faith-based agencies, Visiting Nurse Association and hospice programs.

Check at your MDA office and clinic for other local supports.

National and state organizations

Check with the Area Agency on Aging or the state department of social services to learn if your state will provide funds for hiring and paying a family member as a caregiver.

U.S. Department of Health and Human Services
www.hhs.gov/od
www.cms.hhs.gov/MedicaidStWaivProgDemoPGI/
Offers information about state Offices on Disability.

Family Caregiver Alliance
(800) 445-8106
caregiver.org
Lists state-funded services for family caregivers

National Conference of State Legislatures'
Family Caregiver Support: State Facts at a Glance
ncls.org/programs/health/forum/caregiversupport.htm

National Family Caregivers Association
thefamilycaregiver.org

Personalized Web pages

CarePages
(888) 852-5521
carepages.com

CaringBridge
(651) 452-7940
caringbridge.org

MDA's *myMuscleTeam* care coordination service
mda.org/myMuscleTeam

Finding volunteers

Faith in Action
(877) 324-8411
finationalnetwork.org
More than 1,000 interfaith volunteer caregiving programs across the country.

Caring for elders

The Eldercare 911 Question and Answer Book, by Susan Beerman and Judith Rappaport-Musson, Prometheus Books, 2005

American Association of Retired Persons
(800) 424-3410, (888) 687-2277
aarp.org

Children of Aging Parents
(800) 227-7297
caps4caregivers.org

Indian Health Service Elder Care
ihs.gov/MedicalPrograms/ElderCare

National Association of Professional Geriatric Care Managers
(520) 881-8008
caremanager.org

U.S. Administration on Aging (AoA)
(800) 677-1116
eldercare.gov

National Association of Area Agencies on Aging
n4a.org

U.S. Department of Veterans Affairs Office of Geriatrics and Extended Care
(877) 222-8387
va.gov/geriatrics/

Training for family caregivers

Community-based resources may offer training and classes for family caregivers. Check with local hospitals, home care agencies, Area Agency on Aging, voluntary health agencies, and county and state departments of health.

American Association of Homes and Services for the Aging
aahsa.org

American Red Cross
(800) 733-2767
redcross.org

National Family Caregivers Association
(800) 896-3650
thefamilycaregiver.org

Nurses In Service Training
living-with-als.org/Diane

Tips for Nursing ALS Patients
march-of-faces.org/BBB/2007/hp-2-lp.htm

Training Program and Resources
mediecta.com

Offers training programs, books and Home Care Companion video collection.

Hiring caregivers

"Is Your PCA Driving You Crazy?" Quest, March-April 2007

"The Working Caregiver Dilemma," MDA/ALS News-magazine, November-December 2006

"Despite the Challenge, People Love Their Live-Ins," Quest, November-December 2005

The Personal Care Attendant Guide: The Art of Finding, Keeping, or Being One, by Katie Rodriguez Banister, Demos Medical Publishing, 2007

Caregivers and Personal Assistants, by Alfred H. DeGraff, Saratoga Access Publications, 2002

Avoiding Attendants From Hell: A Practical Guide to Finding, Hiring & Keeping Personal Care Attendants, by June Price, Science & Humanities Press, 1998

For criminal background check:
casanet.org

Asking for help from friends and family

"ALS in a Small Town: Problems & Solutions," MDA/ALS News-magazine, February 2007

"A New Life With ALS: Denial — Don't Deny Yourself Support," MDA/ALS News-magazine, November 2003

"Caregivers: Say 'Yes' to Offers of Help," MDA/ALS News-magazine, August 2002

Organizing volunteers
myals.org/whereicanhelp

Long-distance caregiving

"Handbook for Long-Distance Caregivers"
Family Caregiver Alliance
(800) 445-8106
caregiver.org

'Share the Care' circles

"Share the Care Circles: Keeping the Circle Unbroken," MDA/ALS News-magazine, July 2003

"Share the Care Circles: Who's Going to Do What?" MDA/ALS News-magazine, June 2003

"Share the Care Circles: Asking for Help Can Be the Hardest Part," MDA/ALS News-magazine, May 2003

"It Takes a Village to Fight the System," MDA/ALS News-magazine, September 2002

"ALS Prayer Chain Offers Caregivers a Way to Fight," MDA/ALS News-magazine, June 2002

"Team Up to Share Caregiving," MDA/ALS News-magazine, October 2000

Share the Care: How to Organize a Group to Care for Someone Who is Seriously Ill, by Cappy Capossela and Sheila Warnock, Simon and Schuster Adult Publishing Group, 2004

eHope
(207) 856-7340
ehope.nu

Lotsa Helping Hands
lotsahelpinghands.com

Share the Care
(646) 467-8097
sharethecare.org

Finding or adapting a home

See “Accessibility at Home” resources, page 32.

A contractor’s organization may donate labor; also look for a local Home Builders Foundation.

Medical help

“Social Workers Offer a Wealth of Resources,” MDA/ALS Newsmagazine, September 2007

“How a Nurse Practitioner Fits into the Health Care Puzzle,” MDA/ALS Newsmagazine, November-December 2006

“ALS Case Manager Touches the Lives of Many,” MDA/ALS Newsmagazine, February-March 2005

“Support, Empowerment Keys to Social Worker’s Role at MDA/ALS Center,” MDA/ALS Newsmagazine, June 2003

101 Ways to the Best Medical Care, Charlotte E. Thompson, M.D., Infinity Publishing, 2006

Hospital Survival Guide: 100+ Ways to Make Your Hospital Stay Safe and Comfortable, by David Sherer, M.D., and Maryann Karinch, Adler & Robin Books, 2003

Get Palliative Care
(212) 201-2670
getpalliativecare.org

National Association of Hospital Hospitality Houses (NAHHH)
(800) 542-9730
nahhh.org

NAHHH represents organizations that provide lodging and service for families receiving medical care away from home.

National Patient Travel Center
(800) 296-1217

PatientTravel.org
Helps locate air transportation for patients who need distant specialized medical care.

Patient Advocate Foundation
(800) 532-5274
patientadvocate.org

Visiting Nurse Associations of America
(202) 384-1420
vnaa.org

Long-term care

Long-Term Care, by James L. Matthews, NOLO, 2004

Love, Honor, and Value, by Suzanne Geffen Mintz, Capital Books, 2002

Assisted Living Federation of America
(703) 894-1805
alfa.org

Medicare — “Types of Long-Term Care”
medicare.gov/LongTermCare/Static/HomeCare.asp

National Clearinghouse for Long-Term Care Information
U.S. Department of Health and Human Services
longtermcare.gov

U.S. Administration on Aging (AoA)
(800) 677-1116
eldercare.gov

Hospice

“Research Links Hospice Care With Prolonged Life,” MDA/ALS Newsmagazine, July-August 2007

“Hospice: Comfort and Care,” MDA/ALS Newsmagazine, November 2003

American Hospice Foundation
(202) 223-0204
americanhospice.org

[HospiceDirectory.org](http://hospicedirectory.org)
(800) 868-5171
hospicedirectory.org

Hospice Foundation of America
(800) 854-3402
hospicefoundation.org

Hospice Net
hospicenet.org

Hospice Patients Alliance
(616) 866-9127
hospicepatients.org

National Hospice and Palliative Care Organization
(800) 658-8898
nhpco.org

Assisted living facilities

Consumer Consortium on Assisted Living
(703) 533-8121
ccal.org

National Center for Assisted Living
(202) 842.4444
ncal.org

New LifeStyles
(800) 869-9549
NewLifeStyles.com

Nursing homes

“Nursing Homes: A Good Move?” MDA/ALS Newsmagazine, January 2007

Nursing Homes: Getting Good Care There, by Sarah Greene Burger et al., National Citizens’ Coalition for Nursing Home Reform, Impact Publishers, 2001

“ALS Information for Nursing Home Staff”
living-with-als.org/Diane/inservice.html

National Citizens’ Coalition for Nursing Home Reform
(202) 332-2276
nccnhr.org

Respite help

“Respite Care Benefits Both Patient and Caregiver,” MDA/ALS Newsmagazine, November-December 2006

“Caregiver Respite: The Time It Takes to Make Time Is Worth the While,” MDA/ALS Newsmagazine, December 2005

Arch National Respite Network
(703) 256-9578
archrespite.org

Easter Seals
(800) 221-6827
easterseals.com
Easter Seals provides services, including adult day care.

National Adult Day Services Association
(866) 890-7357
nadsa.org

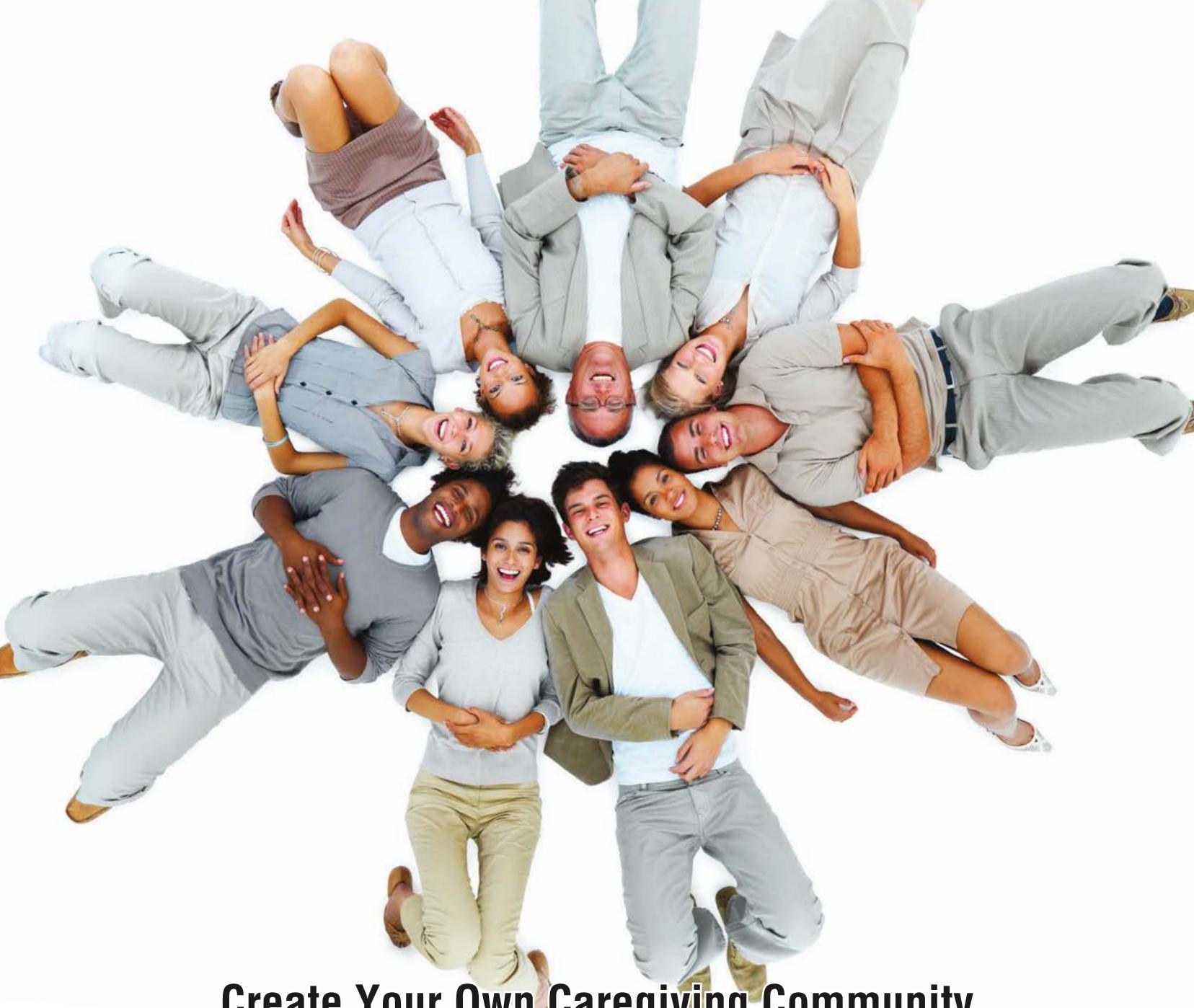
National Respite Locator Service
(800) 473-1727
respitelocator.org

Respite Match (run by a man with ALS)
(678) 884-0281
respitematch.com

Shepherd’s Centers of America
(800) 547-7073
shepherdcenters.org

U.S. Administration on Aging
National Family Caregiver Support Program
AOA National Family Caregiver Support Program
aoa.gov/aoaroot/aoa_programs/hcltc/caregiver/index.aspx

U.S. Department of Veterans Affairs Office of Geriatrics and Extended Care
(877) 222-8387
va.gov/geriatrics
Veterans eligible for outpatient medical services can receive in-home respite care.



Create Your Own Caregiving Community

MDA is proud to offer *myMuscleTeam*, a free and simple way for MDA families to receive strength from a personally created community — their “Muscle Team” of family and friends. Users can create private, secure *myMuscleTeam* Web pages, which offer two primary services.

First, families can post photos and journal entries to keep friends and loved ones updated on medical matters and other aspects of their lives. It’s a great way to update everyone simultaneously, as opposed to the stress-filled and time-consuming process of calling each friend and family member to tell and retell the latest news.

And, families have access to a “care coordination calendar,” where you and/or your primary caregiver can post items for which assistance is needed, such as transportation to medical appointments, meal preparation, household chores and more. That way, family, friends and loved ones can sign up to help. Privacy and security settings enable you to control which family members and friends are permitted access.

For more information and to see how *myMuscleTeam* works,
go to mda.org/services/finding-support/mymuscle-team.

myMuscle Team®

Chapter 9

End-of-Life Issues



NOTE: This chapter should be read only by those who are ready to explore issues related to death.

Saying Goodbye

Most families affected by ALS will have a long time to prepare for the end of the journey. No one knows just how long a person with ALS will survive after diagnosis — in some cases it's only a year or so; in other cases, it's decades. But as the person grows extremely weak, breathing becomes more difficult and quality of life declines, the reality grows closer.

Practical preparations

Before the last stages of the loved one's life, families should have created a list of all checking, savings, investment and other accounts, storage facilities and other properties, wills, legacies and other legal documents. Wills, arrangements for child custody, living wills and powers of attorney should have been taken care of (see Chapter 7). Taking care of these earlier will be easier than waiting until death seems imminent.

Review these documents a few times a year to be sure they're current and your loved one's wishes haven't changed. Also create a list of the people who should be contacted when the person passes. Hospice staff or social workers can check into state laws or local regulations that may affect your plans.

Talking with your loved one about the end of life

Chapter 7 describes the plans to be made and documents to be created regarding medical intervention in times of crisis. The living will should advise doctors that your loved one maintains the right to refuse or withdraw treatment, including mechanical ventilation.

If ventilation is withdrawn, doctors should provide access to adequate pain medications (including morphine) and anti-anxiety drugs. When the day arrives, the person can experience the last moments in some comfort, with dearest loved ones nearby.

“ What your dad most wants to hear from you and the others who love him, is that:

1. He is entitled to make his own choices regarding the interventions used or refused.
2. No matter what he chooses, or his reasons for doing so, you plan to be right there beside him all the way!
3. Nothing can happen to make you not love him more every day.
4. He is a gift to you and your family, and every day with him is an added blessing.
5. You will do everything in your power to be sure he is comfortable and not suffering. ”

“ I am appreciating the ‘long good-bye’ this disease is giving me ... It sure is giving me time to ease the minds of my loved ones, some of them anyway. We’re all learning that dying is a natural part of life. Having been through a few instant ‘no good-byes’ I’d say this is better. ”

“ I did something that, surprisingly, has brought me great comfort. I actually reserved a cemetery plot in an exceedingly beautiful cemetery. It truly looks more like a park than a cemetery and I actually feel so happy to know that where I’ll eventually end up is such an incredibly beautiful place! Next I’ll pick out a marker and have it decorated just as I like with just the right words on it. To me this is not morbid — good planning is a powerful coping tool! ”

There are other questions to think about in advance. Focus on these as they come up naturally in conversation, or initiate a discussion when the time seems right. Remind the loved one that everyone needs to make end-of-life arrangements well in advance and that you want to be sure you know what he or she wants.

Saying Kaddish

by Elizabeth D. Koozmin

I am feeling pretty morose tonight. Today was the last day to say Kaddish for my father, who had ALS, in the Jewish year of mourning.

The first week after death, you stay home and receive visitors, talk about the deceased, and pray at home and recite the Kaddish prayer; the prayer is not about death, it reaffirms your faith. The idea is to have your community come to comfort you and almost never leave you alone. For 11 months after death, you transition slowly back to your normal routines, but you are obliged to recite the Kaddish prayer at every opportunity, to reaffirm your faith in the shadow of tragedy.

So that's what has gotten me through the past 11 months — the routine of going to services at my synagogue (where routine and stability can be

counted on, unlike the rest of my crazy life) and reciting the Kaddish prayer with other mourners. It's supposedly a tried-and-true method of grieving that Jews have been practicing for centuries.

For the 12th month before the anniversary of his death in December, Kaddish is not recited. We will have an unveiling of his headstone, which he shares with my mother who died nine years ago and only six days apart on the English calendar. I will get up and say Kaddish once again in December, one week for him, the next week for my mother.

And with that, the year of mourning will be complete. I am not looking forward to its end, it came way too fast. The years and years ahead without my parents yawn like a big gaping hole that I don't quite know how to fill. Everyone loses both parents eventually but I am only 48 years old with a 13-year-old daughter (who now has no grandparents) and I feel so cheated, selfish, like a small child. I know my husband and I will just have to reinvent family traditions for our daughter's sake.

- Would the person with ALS like to donate tissue to ALS research?
- Would he or she like to donate viable organs for transplant?
- Does the caregiver need to hear that it's OK, at some point, to remarry?
- Legacies for children or grandchildren, such as scrapbooks or taped messages, should be completed and presented to a designated person to save or distribute (see Chapter 6).
- Memorial donations or bequests to charities or hospitals should be included in the will so the estate executor can simply carry them out.

Select a mortuary in advance. Some people with ALS want to be involved in planning their funerals, cremations or burials. Bring this up when the person is speaking about dying, or take note of any mention made in conversation. Some people find a sense of peace and control in choosing a burial plot and headstone, or specifying details of a memorial service;

others are happy to leave that to the loved one. Plans can be paid for ahead of time.

The person with ALS may want to designate clothes for the burial, a list of possessions to give to others, music or readings for the service. Be sure to have biographical details for the newspaper obituary, such as date and place of birth; names and cities of children; primary job titles; major achievements and activities; and where to send memorial donations.

Advance planning will ease the burden on the caregiver at a difficult time. It also can help minimize conflicts with family members. The loved one even can sign a document describing the chosen final arrangements, and there will be less disagreement over what he or she would have wanted.

Keep in mind that funerals or memorial services exist to help the living as they mourn; be sensitive to the needs of close family members at the time of death.

When making final plans, it might be wise to make arrangements for the caregiver as well. In the event

of an unexpected tragedy, the spouse with ALS won't have to deal with those details.

Some people have last wishes. Friends, family or organizations may be able to help arrange for a trip, dinner at a favorite restaurant, a stroll through a favorite park, a concert, a special visitor. Some last wishes, such as taking an airplane ride or meeting a favorite sports team, should be done while the person is still strong enough to get around without danger.

Remember that no one can say for sure when your loved one will pass away. Their remaining time may be more or less than the doctor predicts.

With all of the practical arrangements in place, you can spend your last days or months together in quiet intimacy. Be loving and gentle, say anything that needs to be said, help each other accept the inevitable.

Rites and rituals for the end of life

Memorial services can be traditional and follow the pattern common to the family's religious community, or they can be individual. One person created a DVD with photos, music and favorite quotes to play at the memorial service. Others include special pets, favorite flowers or pictures, candlelight rituals or comments from key people in the loved one's life. One man set aside money for a party at his home with his favorite foods to be enjoyed by all those he cared about.

See "Resources," page 181, for books and websites that suggest memorial rituals that may be particularly meaningful to a family. A ceremony in nature, using water, pebbles, flowers or leaves, may help emphasize the natural cycle of life. The choices are infinite. Look for what has special meaning to you and your loved one.

A memorial gift to the church or community or donating plants to a favorite garden in his or her name can be a fine tribute that helps in the grief process while honoring the person. Carrying out a pet project that had meaning to your loved one with ALS may help to ease the pain of the next months.

Ask people to contribute their thoughts on a website over several weeks. These later could be gathered into a book for close friends and family.

“When [my husband] died I did a DVD photo/music tribute for the memorial service, and many/most of the photos are from after he got ALS.**”**

“On a very special loving evening, he told me what kind of memorial service he wanted, that he hoped friends could come to the house, rather than a funeral home, to pay their respects. He even told me what kind of music to play at the service. I will never listen to that piece again without thinking of him.**”**

These events not only honor the loved one; they help everyone experience grief and appreciate the ongoing nature of life. They can be especially helpful for children and bring everyone to a more meaningful understanding of the person's life.

Grieving the loss

Those who've made the ALS journey as caregivers have much experience with grief (see Chapter 6). There's grief from the time of diagnosis until long after the death. Grieving a major loss lasts a lifetime; most people say they simply get better at coping with it.

See "Resources" for books and other materials on the stages of grief: denial, anger, bargaining, depression and acceptance. Another view of the stages is: numbing, yearning or searching, disorganization or despair, and reorganization.

Separation by death may produce behaviors and emotions indicating that a strong attachment has been broken, including disturbed sleep, forgetfulness, angry outbursts, social withdrawal, feelings of helplessness and dependence, lack of motivation, low self-esteem or anxiety about one's own mortality. There may be physical sensations of grief such as headaches, oversensitivity to light, fatigue, abdominal pain, reluctance to eat, breathing difficulties. Pre-existing conditions may be worsened as immune defenses are lowered.

“This will be my first Christmas Eve without him. I planned to be alone, put a little of his aftershave on my pillow and cry all night!

Well, you know what they say about best laid plans. My 6-year-old grandson has changed that. He will be here and has planned our evening. We will go to church and thank God that we had Papa as long as we did, and he will spend the night in my bed and we will wear Papa's PJs.

I have decided that he is right: We shall celebrate and enjoy all our memories. **”**

“Get out and do things before you feel like it. I promise, the motivation to do things will return much sooner if you will go ahead and do them. Get in motion ... the emotion will follow along naturally. Maybe not as soon as you would like, but it will get there. **”**

Longtime caregivers often feel at loose ends, not sure who they are or what to do with their time, now that the all-consuming caregiver role is over.

If these feelings and sensations don't arise, that's normal too. Anticipatory grief — knowing the loved one is dying and grieving during a series of losses — may reduce the intensity and duration of grief after the death. Feelings of relief also are normal, despite the sadness of loss.

Whatever your emotions or thoughts, allow yourself to feel them and to express your bereavement in your own way. The pain will ease, though not quickly. Watch for signs of unresolved depression or anger (see Chapter 6) and get the help you need.

Friends and relatives may visit and call often at first but become scarce a few days or weeks after the funeral. Call them and tell them you need to talk or to go for a walk together. People never feel adequate in putting their sympathy into words, but that doesn't mean it's insincere. Every gesture of sympathy is a showing of love, so try to receive it that way.

Remember that everyone grieves differently, including each member of the family. Respect each other's processes, and be kind and gentle. Talk with each other about your feelings and listen; don't judge or compare.

Some people hold onto a special talisman of the departed one. A woman kept the headrest from her husband's wheelchair; another slept on the sheets from her partner's deathbed — the last thing he touched on earth. These comfort symbols may carry the person's scent, which is the last strong evidence of physical presence. Holding onto such items is normal, and can be continued for weeks and months if needed. They may bring both comfort and sadness; that's the nature of grief.

There's no rush to removing the loved one's belongings or clothing or to rearrange the household that's been set up for him. The caregiver and family can do this whenever they're ready.

The grieving process isn't a smooth road. Just when it seems you're moving along, something — a familiar song, a person who looks like your loved one, someone else's illness — may bring up the feelings of loss again. There's no set time for grieving — it takes as long as it takes. Anniversaries of birthdays, weddings, the date of diagnosis, and the death itself can intensify or reawaken the feelings of grief. Acknowledge the experience and the emotions, and let yourself seek support.

Take comfort in knowing that the pain is over for the person you loved. Many caregivers imagine their loved ones released from their broken bodies, whole, healthy and happy again.

Also take pride in knowing that you did the best you could, and loved the best you could. If you feel a sense of relief that the hard work of caregiving is over, don't feel guilty. It's been a hard job, and you've earned a rest. Caregivers often know that their loved ones would want them to be happy and to enjoy the relief from the tasks they've performed for so many months or years. Extra care and attention are appropriate right now; it's OK to treat yourself to a massage or manicure, or to go on a trip, or laugh or dance. When you're ready, you deserve to experience the joys of life.

Take care of yourself physically during the weeks and months after the loss. Your immune system will be weakened by the stress of grief, so rest, nutrition and exercise are especially important. Your health can be affected by caregiver stress up to three years after the caregiving burden ends.

Beware of excessively numbing pain with medications, alcohol, television or activity. The grief won't go away, and these escapes can lead to new problems. Professional counseling can help you face the feelings you're avoiding.

Contact MDA for other materials and sources of support.

Support groups and bereavement groups can be especially helpful now. The friends you make through ALS support groups will understand better than anyone what your journey has been and what the loss means. Bereavement groups are offered by hospitals, churches and other facilities. There also are virtual online groups such as griefnet.org. These groups can remind you that your feelings are normal and you're not alone.

When you're ready, consider donating some equipment (such as wheelchairs, communication devices or hospital beds) to your local MDA equipment program, to be shared with other families living with MDA. Think about staying involved with the MDA ALS support group — your experience and perspective can be a tremendous gift to other families. You even might choose to provide respite care, get involved in raising public awareness about ALS or caregiver needs, or volunteer at a hospice or hospital.

As your life goes on, you'll incorporate your loved one's memory into your new experiences. On family occasions and holidays, be sure to include thoughts or mentions of the one who's gone.

Your loved one with ALS will continue to live in your memories and in your heart.

“I thank God every day for the memories of the tiniest caring rituals and love for my husband in the seven years of his journey. So many memories of tweaking his toes, rubbing his leg, combing his hair. Each is a gift life gave me with my most precious husband. Those memories of the care and love help me now that I have to live without him. He is always with me, though. **”**

“Some days are peaceful and we cope. Some days we each seek a corner of the house. **”**

“Recently I've had two occasions where I could have sworn I heard his bed noise when we would turn him. The noise the mattress would make. The bed is not here anymore but I swear I heard the noise. I sat up straight and held on to that moment. **”**

“I did feel relief. Relief that my wife was whole again. Relief from the oppressive burden of the constant care she required. Relief from the daily uncertainty about how much longer she would live. For me, the relief was like a release. Releasing me to begin my healing. **”**

Confessions of a Recovering Caregiver

by Daphne Simpkins

Since my three-year stint as my father's caregiver, I wrestle with socially unacceptable urges to comfort, feed and water just about anybody. I do not have to know you personally to offer you a cough drop when you choke. I say "Bless you" before you finish sneezing, and my right hand will automatically fidget for an Aloe-enriched, bacteria-killing tissue.

After your third sneeze, I will tell you the names of cold products you need, although these medicines are not what I think truly promote healing. Sick people need to go to bed and rest and drink plenty of fluids and be waited on by people like me.

I am ready to do that. I am a recovering caregiver always on the lookout for someone who needs caregiving — me. Sometimes I tire of hearing myself, but I cannot stop. That caregiver urge! I overflow with it.

On an idling airport shuttle bus the other day, the driver asked passengers if we would be responsible to not let another person on if he left the doors open so we could have fresh air. Other passengers nodded politely. I got excited for no one believes in the benefits of fresh air more than a recovering caregiver.

I watched hard. Two people got on. I asked the lady beside me, "What are we supposed to do now?" "It's not our job to guard that door," she said, shrugging.

My jaw dropped. I was envious of that shrug for I have lost track of the boundaries of socially acceptable helpfulness, and I know it. I am labeled by others as codependent, hypervigilant and addicted — one of those suckers born every minute.

But I wasn't born in a minute. My condition evolved over time while I handled medical emergencies for a dying man and forgot who I was, except as a caregiver. I have emerged from that experience in hyper-helpful mode. I watch. I warn. I offer. I am a recovering caregiver and there's no 12-step program to rehabilitate me.

But you could. And you could help others like me or who may become like me. First, you have to see caregivers. They live and move among you, but are very adept at being invisible. To find one, simply look.

Beside a chronic patient is a barely alive, almost-invisible caregiver. Speak to him. To her. Speak these words slowly: "How are you?" If she replies, "fine," smile reassuringly. Send fresh fruit to her house anyway. Or maybe a fresh flower. Drop off fresh milk. Fresh bread. Her life is mostly stale, and she can't easily drive to a store for fresh stuff. You get the idea.

Any gesture or gift of care for a current caregiver who has forgotten her own needs will become a potent memory that will surface later like medicine from a dissolving gel capsule that releases a healing dose of self-recognition and the restorative message: It's okay to accept help rather than only give it.

But don't over-react. If a recovering caregiver you know is already loose and roaming around compulsively offering Band-aids, water, cough drops, and tissues, don't resist them. Instead, simply accept everything a former caregiver offers, and say, "Thank you!"

Caregivers haven't heard those words in ages. Rather than feed an addiction for approval, which some experts warn is what makes caregivers who they are, that expression of simple courtesy will help a caregiver exhale and finally say to someone, "You're so very welcome." The job is done then. See? She is finished. He can let go. Say good-bye.

I know. Every time I say those words, I say good-bye to my old caregiver self and breathe hello to the people who live in the world where I can imagine being on a shuttle bus sitting near a just-about-to-sneeze, almost-gonna-cough, possibly thirsty person, and — oh, bliss — simply shrug.

*Daphne Simpkins is the author of *The Long Good Night* (William B. Eerdmans Publishing Company, 2003), a family memoir about caregiving.*

(Excerpted with permission from Today's Caregiver, caregiver.com)

Resources

Back issues of MDA publications such as *Quest* and the *MDA/ALS Newsmagazine* can be found online at mda.org/publications, or by calling MDA Publications (800-572-1717).

Talking with your loved one about the end of life

Final Journeys: A Practical Guide for Bringing Care and Comfort at the End of Life, by Maggie Callanan, Random House, 2009

Living Fully in the Shadow of Death: Assurance and Guidance to Finish Well, by Susan Zonnebelt-Smeenge and Robert C. DeVries, Baker Books, 2004

Never Too Young to Know: Death in Children's Lives, by Phyllis Rolfe Silverman, Oxford University Press, 1999

Final Gifts: Understanding the Special Awareness, Needs and Communications of the Dying, by Maggie Callanan & Patricia Kelley, Bantam Books, 1997

Twilight Wish Foundation
(877) 893-9474
twilightwish.org

Grants wishes and plans special group celebrations for people 68 or over with low income. Active in several states.

Rites and rituals for the end of life

Crossing the Bridge: Creating Ceremonies for Grieving and Healing from Life's Losses, by Sydney Barbara Metrick, Apocryphile Press, 2006

The Memorial Rituals Book for Healing and Hope, ed. by Ann Marie Putter, Baywood, 1997

Grieving the loss

"Following Sam: Fond Farewell," MDA/ALS Newsmagazine, February 2008

Coping With Grief: Strategies for People Living with ALS, ALS Society of Canada, als.ca/en/publications-and-resources/coping-grief

On Grief and Grieving, by Elisabeth Kübler-Ross, Scribner, 2005

Parting: A Handbook for Spiritual Care Near the End of Life, by Jennifer Sutton Holder, University of North Carolina Press, 2004

When Bad Things Happen to Good People, by Harold S. Kushner, Schocken Books, 2004

Tuesdays with Morrie, by Mitch Albom, Random House, 2002

Elisabeth Kubler-Ross Foundation
ekrfoundation.org

Organization created to further the work and teachings of Elisabeth Kubler-Ross, particularly her work on behalf of improving end-of-life care.

GriefNet
griefnet.org

Resources for children

Tear Soup, by Pat Schweibert and others, Grief Watch, 2005

The Fall of Freddie the Leaf, by Leo Buscaglia, 20th anniversary edition, Slack, 200

The Gift of a Memory: A Keepsake to Commemorate the Loss of a Loved One, by Marianne Richmond, Marianne Richmond Studios, 2001

Guiding Your Child Through Grief, by James P. Emswiler, Bantam, 2000

Dougy Center for Grieving Children and Families
dougy.org

Hospice Net
hospicenet.org/html/child.html
KidsAid.com (produced by GriefNet)

Appendix A

MDA's Site *myMuscleTeam* Helps You Keep in Touch and Coordinate Volunteers



In the journey with ALS, a good support network is essential. Friends, family and willing volunteers make life easier and sweeter in so many ways.

But coordinating such a network can seem like one more job on an overflowing “to do” list. It takes time and effort to keep people informed and to connect vague offers of help to actual tasks.

To address this problem, MDA offers *myMuscleTeam*, a free online service that helps MDA families experience the strength and support that comes from their community — or “Muscle Team” — of friends and family.

The site allows individuals with ALS and their family/caregivers to create private and secure Web pages that serve two primary functions: communication and care coordination.

Communication: On *myMuscleTeam*, users can post photos and journal entries that keep their support network updated on medical matters and life in general. Users only have to post once to reach multiple recipients, as opposed to telling and retelling the latest news to separate individuals. Because the site automatically sends out a notice whenever an update is posted, it also is helpful to those who worry about “bothering you” with questions, but who want to stay up-to-date and aware of your changing circumstances. The end result: Your support network grows stronger with less effort on your part.

Care coordination: A care coordination calendar enables users to post items for which assistance is needed — such as transportation to medical appointments, meal preparation, shopping, household chores and more — so

family, friends and others from your personal “Muscle Team” can sign up to help. Privacy and security settings ensure that you control which people are permitted access to the calendar. An automatic reminder email is sent to those who have signed up to help. This calendar is the perfect answer to the oft-asked question “What can I do to help?”

As you look ahead, take a moment to check out *myMuscleTeam* at mda.org/mymuscleteam, and put it to work for you!

Reprinted from the MDA/ALS Newsmagazine, January-February 2010



Appendix B

When the Thinking Parts of the Brain Go Awry in ALS

by Amy Labbe

John Shearer and his wife Brenda learned in November 2010 that he has ALS. The couple, who lives in Woodstock, Ga., was told the disease would cause gradual weakening of John's voluntary muscles, such as those used for moving and breathing.

John, 70, retired four months later in March 2011 from Lockheed Martin Aeronautics, where he worked 41 years as a machinist.

As predicted, over the past year the muscles in John's legs and hands have weakened, and he now needs noninvasive ventilation at night to help him breathe.

But that's not all. Although John is still able to talk, Brenda says, he "doesn't carry on conversations anymore."

This behavior change is due not to muscle weakness, but to John's difficulty in processing words, focusing and paying attention, following what's being said, and formulating an appropriate response, Brenda says. She likens his symptoms to a computer with a faulty processor that, when it works, does so with a slow modem.

"When I ask John something, it's like it just doesn't register," she says. "And sometimes he never answers."



Cognitive changes in ALS

Traditionally, ALS has been described as a neurodegenerative disease affecting the motor neurons that control muscles. Involvement of other "non-motor" systems was thought to be rare.

But cognitive and associated behavioral symptoms associated with ALS have been noted in studies dating as far back as 1889. Today, it is known that approximately 50 percent of the time, ALS is accompanied by some degree of frontotemporal lobar degeneration (FTLD), a type of progressive deterioration in the frontal and temporal lobes of the brain (situated in the area of the forehead and extending approximately as far back as the ears).

ALS is now known to be a multisystem disorder. Loss of motor neurons still is the hallmark of the disease, but non-motor involvement, including degeneration in parts of the brain responsible for cognitive function, also is recognized.

Although roughly half of all people with ALS exhibit at least some symptoms of cognitive impairment at some stage of their disease, only a small percentage are like John Shearer, whose severe cognitive and behavioral difficulties meet the criteria for an official diagnosis of frontotemporal dementia (FTD).

Usually, cognitive and behavioral symptoms in ALS range from mild (such that only close family members may notice a difference) to moderate. Symptoms include atypical behaviors, apathy and poor judgment, anger and irritability.

The simultaneous occurrence of both disorders is called ALS-FTD.

An overlap between ALS and FTD

These two distinct disorders — FTD and ALS — are merging, and scientists are coming to understand that there's tremendous overlap between the two, says Stanley H. Appel, chairman of the Department of Neurology at the Methodist Neurological Institute, in Houston, and director of the MDA/ALS clinic there.

Scientists have identified several genes that appear to be associated with both ALS and FTD. In the fall of 2011, researchers uncovered a gene, C9ORF72, which they identified as the most common cause to date of familial ALS, FTD and ALS-FTD.

In addition, a 2005 study found FTD-associated changes in the brains of all ALS participants, whether or not they showed signs of cognitive impairment.

"We did a study in ALS patients who were cognitively normal, and they had changes in the frontal and temporal regions of the brain consistent with FTD," says Catherine Lomen-Hoerth, who directs the MDA Clinic at the University of California at San Francisco (UCSF) Medical Center. "It's just to a much lesser degree, so there are no obvious symptoms."

Symptoms of FTD in ALS

The term "FTD" in ALS can be confusing. This is because physicians and researchers often use it in a general way to refer to the entire spectrum of cognitive and behavioral problems that may occur, from barely noticeable "mild cognitive impairment" to disturbances of a more profound nature that satisfy the criteria for an official diagnosis of FTD.

Most people's perceptions of "dementia" include the

dramatic memory loss, confusion and disorientation often seen in people with Alzheimer's disease.

In ALS, however, "dementia" does not typically affect memory or render people unable to recognize their loved ones. Dementia associated with ALS typically causes only mild symptoms, often noticeable only by those who know the person well.

The parts of the brain work together via neural networks, explains Michael Strong, dean of the Schulich School of Medicine & Dentistry and professor of neurology at the University of Western Ontario.

Disconnects in these networks due to damage caused by disease can happen anywhere in the brain, he explains. In ALS, they occur in the frontal and temporal lobes governing the higher thought processes that make up "executive function." Such processes include: making or following complicated plans, solving complex problems, following a series of directions and making sound judgments.

Individuals with executive dysfunction may have problems completing tasks that require complex planning, forethought or organization.

In the majority of cases, cognitive changes are mild. But sometimes they can be significant — such as John Shearer's difficulty conversing. "Think of some of the

Cognitive Symptoms in ALS: A Caregiver's Role

Families can deal with the cognitive and behavioral changes associated with frontotemporal dementia (FTD) by making modifications to the environment (i.e., keeping certain items in designated locations, and using calendars or notes to organize information), and by carefully describing all symptoms to health care providers and medical professionals.

It's important to help people with ALS and FTD to make medical care and end-of-life decisions — and legally document them — as early as possible. This helps to avoid difficulties later when they may be physically or legally unable to make such decisions on their own.

Caring for someone with the physical symptoms of ALS is a big job, and when FTD symptoms also are present, caregivers may feel even more overwhelmed. In addition to experiencing chronic depression, caregivers often report feelings of frustration, anger, guilt and anxiety.

Caregivers should not hesitate to speak with their loved one's ALS physician about their situation. They also should seek support from family members and an ALS support group, such as those offered by MDA in local communities, on the telephone and online.

For help locating a support group or to learn about other resources that may ease caregiver stress and burden, contact your local MDA office at (800) 572-1717, or visit mda.org.

more eloquent things that we do, neurologically speaking,” says Strong. “For example, think of speech and language, and how we string a sentence together and have it be smooth, so it’s not hesitant or nonsensical; and how we don’t substitute words inappropriately.”

Behavioral changes can include:

- acting inappropriately in public or toward loved ones and caregivers;
- loss of motivation (apathy);
- diminished recognition and response to the feelings and needs of others;
- repetitive or ritualistic activities or habits; and
- a change in diet that can include new “favorite” foods or eating too much in one sitting.

Some researchers suspect that FTD-related changes in the brain also may underlie pseudobulbar affect (PBA, or emotional lability), although a definitive relationship has not been shown.

PBA, which causes excessive and uncontrolled laughing or crying, can be treated with Nuedexta, a new medication recently approved for use in ALS by the U.S. Food and Drug Administration.

Who gets FTD symptoms?

Approximately half of all people with ALS won’t exhibit any cognitive or behavioral disruptions throughout the course of their disease.

The other half will exhibit some signs of FTD at some stage in their disease. In most cases, the signs will be extremely subtle.

It’s estimated that between 10 to 30 percent of people with ALS will have more severe FTD symptoms.

Studies have shown that older age, symptoms beginning in the speaking or swallowing muscles (bulbar-onset ALS), and a high degree of bulbar involvement correlate with an increased incidence of FTD.

FTD and ALS cause “a wide range of different presentations and severities,” says Walter Bradley, former director of the Kessenich Family MDA/ALS Center at the University of Miami School of Medicine.

“You have people with absolutely pure ALS, who never show any signs of cognitive impairment whatsoever, or any behavioral abnormality, or any other of the hallmarks of frontotemporal degeneration,” says Bradley. “And you have people who have pure frontotemporal dementia, who never get any motor neuron disease, no ALS.”

“And then you get people who are somewhere in between the two ends of that spectrum.”

Testing to diagnose FTD

Lab tests usually aren’t used to make a definitive diagnosis of FTD, although blood or other biological samples may be tested to rule out other factors that can cause dementia-like symptoms.

Anxiety, depression, sleeping problems, insufficient blood-oxygen levels and medication side effects can cause symptoms that mimic FTD-related symptoms, and must be ruled out before an FTD diagnosis can be made.

Imaging studies are useful tools, as they are able to pinpoint brain abnormalities that can be associated with FTD. Computed tomography (CT) scans are able to detect atrophy in the frontal lobe, magnetic resonance imaging (MRI) can help clinicians observe brain abnormalities in finer detail, and single-photon emission computed tomography (SPECT) images are able to indicate brain-function problems.

In 2011, an international consortium developed revised guidelines for the diagnosis of “behavioral variant frontotemporal dementia” — the most common type associated with ALS — based on the available literature and collective experience. The criteria guide physicians toward, or away from, an FTD diagnosis based on the presence and severity of symptoms.

Neuropsychological tests designed to assess different thinking functions (attention, language, memory and others) also can be used to evaluate cognitive changes in people with ALS.

“As far as symptoms go, people can start with ALS symptoms, they can start with FTD-type symptoms, or they can start with both at the same time,” Lomen-Hoerth noted. (John Shearer’s cognitive problems started three years before his ALS diagnosis, Brenda Shearer notes.)

FTD is Not New in ALS

Old descriptions

- Withdrawn due to depression
- Stubborn
- Seeking control in some area of life
- Anger outbursts due to frustration with ALS
- Denial
- Language problems due to dysarthria

FTD-associated behaviors

- Apathetic
- Disinhibited
- Poor judgment
- Easily frustrated
- Quick to anger
- Lack of insight
- Language difficulty
 - Word finding
 - Spelling
 - Aphasia

Cognitive and behavioral changes in ALS used to be described as reactions to the physical symptoms of the disease, such as being “depressed” or “seeking control,” says Catherine Lomen-Hoerth, director of the MDA Clinic at the University of California at San Francisco Medical Center. She says the old phrases used to describe these changes have given way to a list of the behaviors caused by FTD.

Finally, interviews with family or friends also can help medical professionals determine whether someone with ALS is exhibiting signs of FTD, and can, in fact, be the most reliable of all available means of making an assessment.

For example, there are people who are just stubborn, Lomen-Hoerth says. “But what we really look for is a change in behavior from their baseline. So, if someone’s always been stubborn, we’re not going to label that a symptom. But if the family says, ‘no, they didn’t used to be like this,’ then that’s a red flag that something is going on.”

Medical care for people with ALS-FTD

When FTD accompanies ALS, Stan Appel says, it impacts both clinical care and the physician-patient relationship.

People with FTD symptoms can lose insight into their disease process and not realize that they aren’t thinking as clearly as before, or that their behavior may be problematic.

Studies show that only about one-quarter of those with a diagnosis of ALS-FTD comply with recommendations for noninvasive ventilation and feeding tubes, as compared with compliance from three-quarters of those with ALS alone. Perhaps because of noncompliance with such life-supporting interventions, life span tends to be shorter for people with ALS-FTD than for those with ALS alone.

A multidisciplinary ALS clinic is “key” for people with ALS-FTD, says Susan Woolley-Levine, a clinical neuropsychologist at California Pacific Medical Center and the Forbes Norris MDA/ALS Clinic in San Francisco. “The extent of support alone is extremely meaningful to patients and families, but it is also a place to receive more comprehensive care.”

For example, MDA/ALS clinics, which utilize a multidisciplinary approach, offer access to experts such as neurologists, respiratory therapists, physical therapists, occupational therapists, social workers, psychologists, nutritionists, nurses and many others.

This variety of providers can help meet myriad needs, Woolley-Levine notes, from daily living challenges to insurance problems. Clinics also can provide resources and support for caregivers and families.

Knowledge can help

It's important for caregivers to know about possible cognitive symptoms in ALS, so that they can recognize the signs and not think that their loved one is simply "being difficult," "not trying hard enough" or "acting out on purpose."

Brenda Shearer recalls "chewing John out, because he wasn't paying attention to me," and chiding him about "leaving if you don't start paying attention" — until she learned what was actually happening. "I thought he was doing it on purpose," she says. "I didn't know he was sick."

"Patients often don't recognize the changes in their behavior, and often will say, 'Well, there's absolutely nothing wrong with me,'" Lomen-Hoerth explains. "But the families will be nodding and saying, 'Well, actually, we've noticed some things.'"

Letting families know that changes in behavior are due to the disease often brings relief.

"Once caregivers understand that these cognitive and physical changes are not intentional, it helps them to be more patient and to not take the changes personally," says Woolley-Levine.

Michael Strong adds, "To have a physician say there's a disease causing these disturbances changes everything, because now families don't think it's their fault. They know it's part and parcel of the FTD."

For more information:

["Research Roundup: C9ORF72 mutation is most common cause of familial ALS, FTD," MDA/ALS Newsmagazine, November-December 2011](#)

[Video from MDA/ALS Clinic Director Stanley Appel discussing cognitive changes in ALS,](#)

alsn.mda.org/article/when-thinking-parts-brain-go-awry-als#Appel_video

Reprinted from the MDA/ALS Newsmagazine, November-December 2011

Appendix C

Talking To Your Kids About ALS

by Aimee Chamernik

As we huddle on the couch, munching popcorn and engrossed in our movie, the kids and I are startled by a sharp knock at the door. Before I can shift forward to start the painstaking process of standing up, our neighbors burst into our house.

"Are you OK?!" Beth asks breathlessly.

"Yes, we're fine," I answer sheepishly, as it dawns on me that I never called my husband back after Nick left a message for him telling him I'd fallen. I'd been so preoccupied with reassuring Nick (9), Emily (7) and Zachary (3) that I was OK — and responding to Zachary's persistent demands for popcorn — I'd forgotten to follow up on Nick's frantic call. "I'm so sorry!"

"Oh, we're just glad you're OK!" they say in unison. "Jim called because your phone is off the hook, and he was worried."

I dig the phone out of the couch cushions and click "end."

"Oops! No, we're fine. I'll call Jim right away."

The kids, at first too stunned to speak, start spouting a hundred questions at once.

"What are you doing here?" "Did you think our mom was hurt?" "How did you know she fell?"

After a few moments of small talk — apologies and profuse thanks on my side, gracious reassurances on theirs — they pull the door shut behind them.

"Well," I say, turning to the kids, "I feel terrible that we made everyone worry, but that was kind of good practice for an emergency."

"Like a tornado drill at school," Emily nods.

"At least we know we can count on our neighbors for help!" Nick pipes up.

And those words make this embarrassing incident worthwhile. I'm glad he feels reassured.

Since ALS has invaded our lives, the toughest challenge has been embroiling our innocent children in this losing



Aimee Chamernik, 37, of Grayslake, Ill., and (from left) sons Nick, 9; Zachary, 3; husband Jim and daughter Emily, 7. Chamernik writes about talking with her children about ALS.

battle. While we were tempted to try to shield them from ALS for as long as possible, we've ended up taking the opposite approach.

After two-plus years, do we have all the answers? Hardly. But we're finding that a straightforward approach helps to draw our family closer together through this experience.

Here are some of the tenets we try to live by:

Be honest

We talk honestly about ALS with our kids, answering questions in an age-appropriate way. We've promised that we will always tell them the truth, and if we don't know an answer we will try to find one.

Our conversations have ranged from matter-of-fact to heart-wrenching, but the unifying characteristic is that after all of them the kids feel better. By discussing their questions or fears openly, we can help them deal with their feelings.

Don't dwell on ALS

As my balance and coordination began to fail, we slipped

into the habit of telling the kids to pick up their toys “or Mom could fall.” We quickly realized that we were wrongly shifting the focus to ALS. If ALS weren’t part of our lives, we would still teach our children to pick up their toys.

While ALS is unavoidable at times, we try to steer clear of needlessly emphasizing the disease that already commands so much attention.

Keep persevering

You know how kids have that uncanny knack for never hearing “please put your dirty clothes in the hamper” but always hearing what you’d hoped slipped past them? In the back of my mind is the knowledge that my children are watching and learning from me every moment of every day.

I try to let that knowledge guide my behavior on the days when I want to hide under the covers and never come out. Is that how I want them to remember me dealing with adversity? Or how I want them to face adversity in their lives?

No, I want to teach them to dig deep and find the inner strength to live each moment to the fullest. And I don’t hesitate to talk about that choice.

“This is one of those times when I just want to give up,” I’ll say. “But I will not let ALS win!”

Let the kids help

This one’s tough. I’m struck by how topsy-turvy our lives have become; my kids now tie my shoes instead of having me tie theirs. I struggle with becoming a burden, but they are happy to help. Whether they are fetching things I’ve dropped, carrying my purse or shielding me in public places, they’re glad to be my arms and legs.

“When I keep people from bumping into you, I feel like your knight in shining armor!” Emily grins proudly.

Furthermore, they’re learning patience, kindness, and sensitivity to the needs of others. This isn’t how anyone would choose to instill those traits, but I can’t help but be grateful for that positive.

Take control where we can

Jim and I often lament our lack of control, with ALS constantly lurking and snatching more from our family. We can

only imagine how much stronger that feeling is for the kids.

One way we can take back a measure of control is by trying to make a difference in the fight against ALS. The kids are eager to join fundraising efforts, participate in events to raise awareness, and write or draw about their experiences with ALS.

We also establish some control by staying a few steps ahead of my disease. Whether we’re installing grab bars, removing household hazards, or enlisting neighbors to check on me, we keep the kids involved in our safety measures and plans.

When our neighbors burst in on us that night, the kids talked about it for weeks. They are comforted by the fact we have people who care about us and are ready to help out at a moment’s notice.

And while we’d rather shield the kids from ALS entirely, we all find comfort in facing it together.

Aimee Chamernik received a diagnosis of ALS in 2004.

Reprinted from the MDA/ALS News magazine, April 2007

When a Parent Has ALS

by Amy Labbe

Tips for helping children cope

"ALS affects my life, too!" says 13-year-old Lauren Skager of Minneapolis.

Lauren's mom, Gae, has ALS. Lauren sometimes translates for her, but more and more Gae uses her communication device to speak. Lauren worries people will become impatient waiting for her mom to type out what she wants to say.

"I wish they could understand her like me," Lauren says, noting that she feels "relieved" when her mom finishes typing her message and "an embarrassing incident is averted."

Lauren's worry is but a small example of the myriad ways ALS affects not only the individual but the entire family. To help children live full lives despite ALS worries, check out the following tips from the Skagers and other families touched by the disease.

Participate in a youth support group

Don't overlook one-on-one or family counseling, but consider looking for a child/youth support group as well.

Gae, who worked as a registered nurse prior to receiving her diagnosis, says children can gain significant benefit and support from talking with peers.



Gae and Lauren Skager with their golden retriever puppy, Dakota, encourage people across the nation to organize ALS support groups for children and youth.



Sean Dwyer, top, with father, Pat, and Brenna Dwyer find numerous ways to enjoy the present and focus on the positive things in their lives.



"Because this disease is so rare and devastating in its progression, children often feel isolated and alone in their emotional struggles," she notes.

Peers who also have a parent living with ALS (or other chronic conditions like cancer or multiple sclerosis) can provide insight to each other in coping with family changes.

Lauren participates in MDA's Support Group for Children/Youth of ALS Patients in Minneapolis. Facilitated by a pediatric neurologist and attended by approximately nine children ages 10 to 17, the group meets bimonthly for 90 minutes.

Lauren says she enjoys going to the meetings and finds the sessions "interesting and very informational. It helps to know that other kids are coping with the same situation."

Support doesn't have to be face-to-face; you can find it from the comfort of your living room with a click of the mouse.

As a teenager, Sarah Butler (now 21) of Tucson, Ariz., frequented Yahoo's Living with ALS group. There she posted questions and stories, vented frustrations and made supportive friendships that helped her deal with her father's ALS.



Justin Alderman with sisters Jessica, left, and Breanna, says "it's OK to be a little worried" about a parent with ALS. "But don't let it control you."

MDA offers several ALS-specific chats each week; go to mda.org/chat/calendar.html to see the schedule and sign up. The ALS Chat by ALS Forums can be found at alsforums.com.

Stay busy, have fun

School activities, hobbies, pets and part-time jobs all serve as outlets and touchstones for kids and teens.

Brenna Dwyer, 18, and Sean Dwyer, 16, of Kenmore, Wash., say their activities have helped them deal with their father Pat's ALS.

Brenna, a high-school senior, particularly enjoys horse-back riding. She says her horse, Bailey, always has been her "shrink" and that when she rides she's unable to think of anything else.

"Whenever my mind does wander off, she's smart and sensitive enough to let me know that she's the one I need to be paying attention to," Brenna says.

Sean, a sophomore, plays drums in a band, plays basketball, skis and makes movies of his friends skiing. He also enjoys spending time with his dad rebuilding a vintage Triumph TR-6 convertible.

"What I do to not worry as much is to just live life in the present and take advantage of every moment that I have with my dad," Sean says. "I just try to have so much fun and to be so content that I just forget about all the bad things."

Learn and teach about ALS

Knowledge often inspires a sense of empowerment, and children who learn about ALS find themselves in the unique position of being able to educate others about the disease, both by sharing what they've learned and by demonstrating it's nothing to be embarrassed about.

Breanna Alderman, 17, of South Jordan, Utah, won a trophy in an essay contest when she was in the sixth grade, after writing an essay about her father Alan's courage in facing ALS.

She says at first, however, she was ashamed of ALS and what it was doing to her dad.

"After a while I realized that there was nothing we could do about it, so I might as well learn from it," Breanna says. "So, I try and teach other people about the disability and other [disabilities] like it."

When her friends come over, Breanna tells them it's OK to talk to her dad and explains to them about the disease.

Alan says Breanna and her younger brother and sister, Justin, 15, and Jessica, 11, are quick to stick up for him and for others with disabilities. Most of all, though, he says they show others that ALS is nothing to be embarrassed or ashamed about by continuing to "excel and go on with their lives."

Communicate and engage

Open all lines of communication and stay involved.

Alan Alderman and his wife, Shaun, find support in their extended family and church group, and Alan notes, "When I was first diagnosed we also met with all of our children's teachers, coaches, dance and music instructors, and others that associated with our children to make them aware of the situation and to ask for their help in watching out for our kids."

The Aldermans also have family meetings to discuss Alan's condition, where he says they openly discuss what's going to happen and "encourage their questions."

Alan advises parents, "Don't let ALS be the reason for doing, or not doing, things."

"Continue to be engaged and involved in their lives as much and as long as you can. And love them!"

Reprinted from the MDA/ALS Newsmagazine, March 2008

Having Children After an ALS Diagnosis

by Miriam Davidson

Erin Brady Worsham and her husband tried to have a baby for six years, with no luck. They had accepted they would never be parents. Then, in a twist of fate worthy of a novel, Worsham got pregnant the day after she was diagnosed with ALS.

When Worsham learned of her pregnancy, she was delighted. Her neurologist, however, was not. "Don't you know you could be gone in a year?" he asked.

Since ALS tends to strike later in life, few people face the difficult decision of whether or not to have children after a diagnosis of the disease. For those who do, there are many factors to consider, including the type of ALS, the willingness and ability of the family caregiver, financial considerations and the best interests of the child, among others. Some people in this situation decide not to have children; others, like Erin Worsham, are determined to.

Worsham, of Nashville, Tenn., went in search of a sympathetic doctor. She soon found an obstetrician who told her what she wanted to hear: "It doesn't matter if you live five days, five months or five years, you're going to be happy you had this child."

The second doctor was right. Worsham, 51, has outlived all predictions, and her 15-year-old son Daniel has grown into a strapping young man.



Erin Worsham and son Daniel, who was born nine months after Erin received a diagnosis of ALS in 1994.



Keith Gawrick, wife Yvette, and daughters Laura, Katherine and Sofia. Keith received an ALS diagnosis in 1999.

Many issues to consider

Keith Gawrick, of Katy, Texas, had only been married about six months when he received an ALS diagnosis on January 1, 1999 — his 31st birthday.

"One of our first questions was how this would affect our plans to have a family," Gawrick recalls. He and his wife were relieved to learn that he had the sporadic, and not the familial, form of the disease — meaning the chances of him passing ALS on to his children were slim to none — but they had other concerns as well.

"We considered many different aspects of the illness," Gawrick says. "How long would I be around, how would our young children deal with the progression, would they ever understand what happened to me? After much consideration and praying, we decided to enrich our lives with the joys of having children. Not wanting to give in to the fear of the illness, we went ahead and had our family." The Gawricks now have three girls, ages 9, 8 and 16 months.

Gawrick says his kids keep him smiling, even though there are days when he struggles "with the reminders of not being able to be physically engaged" with them. One of the few good things about having ALS, he notes wryly, is being relieved of diaper duty.

"I share the joys that other parents experience, but I don't know how long I will be around and this thought is always

on my mind as I watch my children grow," he says.

Weakened respiratory and abdominal muscles can pose some special risks for women with ALS during childbirth. But for the Worshams, Gawricks and other couples facing a life-threatening illness, having a child can be a way of ensuring a connection with the future.

"Just knowing I had sperm out there gave me a reason to live" is a sentiment some cancer patients have expressed to researchers in connection with freezing sperm prior to undergoing cancer treatment, says Patricia Hershberger, an assistant professor at the University of Illinois, Chicago College of Nursing, who studies young adult women with cancer and their decisions about egg freezing.

Brian Stanfield, of Tulsa, Okla., and his wife Janine took advantage of similar reproductive technology in their quest to have children. Like Gawrick, Stanfield received a diagnosis of sporadic ALS as a young newlywed. He and Janine decided to start a family right away, in order to give Brian as much time as possible with the kids before his ALS symptoms became too severe. After a year of trying to conceive naturally, the couple turned to in vitro fertilization (IVF).

The IVF was successful. The Stanfields had a son and were able to freeze additional embryos. "This became an important issue when we were ready to try for a second child," explains Janine Stanfield. "Brian was enrolled in a clinical drug trial. Since it was an experimental drug, there was little to no information regarding its possible effects



Brian Stanfield, wife Janine, son Braden and daughter Cayla. Brian received an ALS diagnosis in 2003.

on a fetus. We were able to use our frozen embryos and were blessed with a baby girl."

The Stanfield children are now 5½ and 4, and Janine says the family has its share of struggles, particularly about how to involve an increasingly immobilized Brian in the children's energetic, active lives. These same challenges also are faced by families who had children prior to an ALS diagnosis.

"Brian wants to be that dad who teaches his son to play every sport and throws the kids around in the pool," Janine says. "It breaks his heart when they yell for help, and he can't run to the rescue and scoop them up. We try to focus on all of the time he gets to spend with the kids and everything he CAN do with them, like video games, play with baby dolls and go to the zoo.

"The joy of watching the kids play, hearing their laughter and hearing them say, 'I love you, Daddy!' makes Brian smile every day," Janine continues. "We are firm believers that having a positive attitude goes a long way with this disease and our children have added to that 'positivity' time and time again. Life has been beautiful with them in it, even with this dreaded ALS."

Unappealing and problematic

For some couples, having a child when one person has an illness that's likely to drastically shorten life isn't appealing or turns out to be far more problematic than they had predicted.

Steven Albert, a professor in the Graduate School of Public Health at the University of Pittsburgh, studies health outcomes, decision making and quality of life in aging and chronic disease, including ALS.

He's asked some younger couples with ALS about their childbearing decisions. In one case, he says, a couple in their mid-20s decided not to have children after the man received an ALS diagnosis, apparently because they wanted to devote undivided attention to each other during the limited time remaining for him.

"She said her goal in life now was to spend as much time as possible with him," Albert recalls.

In another case, a couple went ahead with plans to have a child despite the husband's ALS diagnosis and things did not go well, Albert says.

The woman ended up feeling that she had to “shield the child from the illness,” he recalls. “That was very hard on the patient. She was very uncomfortable having a little child exposed to a patient with ALS. She thought it would scar the child in some way, which probably is inappropriate, but that’s how she felt. She was not ready for this sort of thing. They ended up divorcing.”

Stories like this illustrate the need for couples to discuss all the ramifications of their decision before going ahead, says Karen Toennis, a nurse and MDA/ALS Clinic coordinator at Methodist Neurological Institute in Houston. Toennis not only counsels couples in this situation; she herself has faced it.

ALS made bearable

Toennis’ husband received an ALS diagnosis at age 37, when they’d been married barely two years. She calls their choice to have a child “the best decision we ever made,” because being a father gave her husband so much joy. “He said having our son made living with ALS bearable.”

Toennis took the opposite approach of the woman who felt she had to shield her child from the disease.

“Kids need to know the truth, but a simple truth,” she says. “I told [our son] Joe, ‘Daddy’s arms and legs don’t work, but he can understand every word you’re saying and he loves to feel your touch.’ My husband couldn’t move a muscle, but our son didn’t care. He adored his dad.”

Toennis’ husband stayed playful and focused his energy on their child. “When he went on BiPAP [a ventilator], we joked that he was ‘Spaceman Daddy.’ He had an eyegaze computer, and he used it to go shopping for birthday and Christmas presents, to plan parties and family vacations, and to organize our son’s Boy Scout activities. He was as involved as he could be.”

Toennis did shield her son in one way, however. “I didn’t ask Joe to do much caretaking,” she says. “I wanted him to enjoy his dad.”

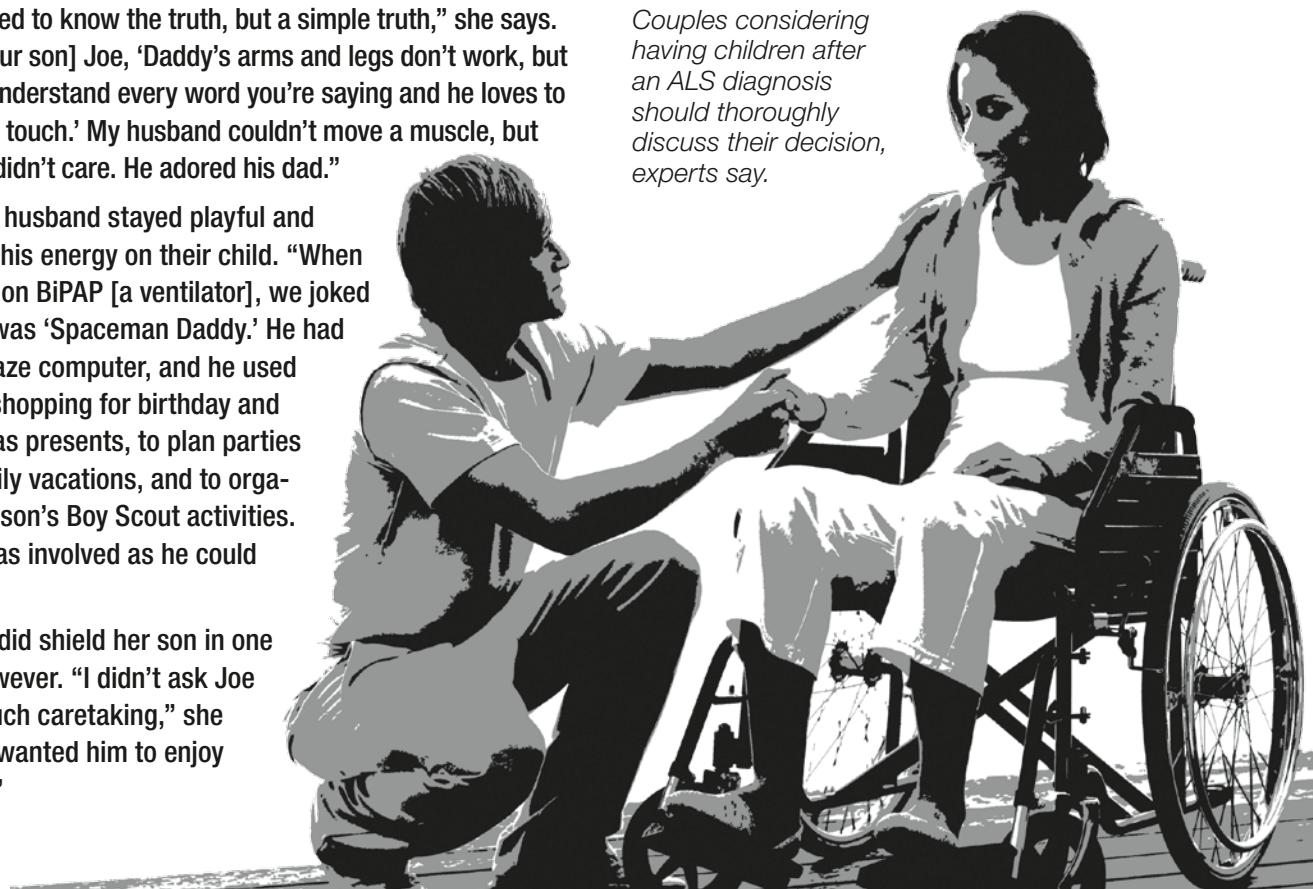
Toennis’ husband passed away in 2006, when their son was 12. She says Joe of course misses his father, but he’s adjusted well. As Toennis points out, they had a good relationship, and Joe got to spend more time with his dad than a lot of kids with fathers who are healthy but absent.

“I tell young couples in the clinic who are considering this that they need to have good support to help take the burden off the caregiver,” Toennis advises. She also points out that people who get ALS at a young age tend to live longer and do better than those who are older when receiving a diagnosis. And for some couples — such as the Worshams, Gawricks and Stanfields — having a child may give a person with ALS incentive to live.

“Everyone goes into having kids not knowing what the future holds,” Toennis says. “In our case, we appreciated every moment together, because we knew we had a limited amount of time.”

Reprinted from the MDA/ALS Newsmagazine, July-August 2010

Couples considering having children after an ALS diagnosis should thoroughly discuss their decision, experts say.



Appendix D

A Closer Look: Diaphragm Pacing System

by Amy Labbe

For Pat Dwyer, this newly approved respiratory assist device means ‘a life better lived,’ but physicians urge caution until more is known

Pat Dwyer of Kenmore, Wash., uses three different devices to make sure he’s getting enough air. A “sip-and-puff” ventilator supplements his breathing efforts during the day; bilevel positive airway pressure (BiPAP) helps him breathe at night; and the NeuRx Diaphragm Pacing System, or DPS, stimulates his diaphragm (the primary breathing muscle) 23 out of every 24 hours.

Dwyer learned he has ALS in June 2005. His symptoms began in his left hand and traveled up his arm; the muscles in his right arm and hand were affected next, and then his diaphragm. He didn’t experience symptoms in his legs until 2009.

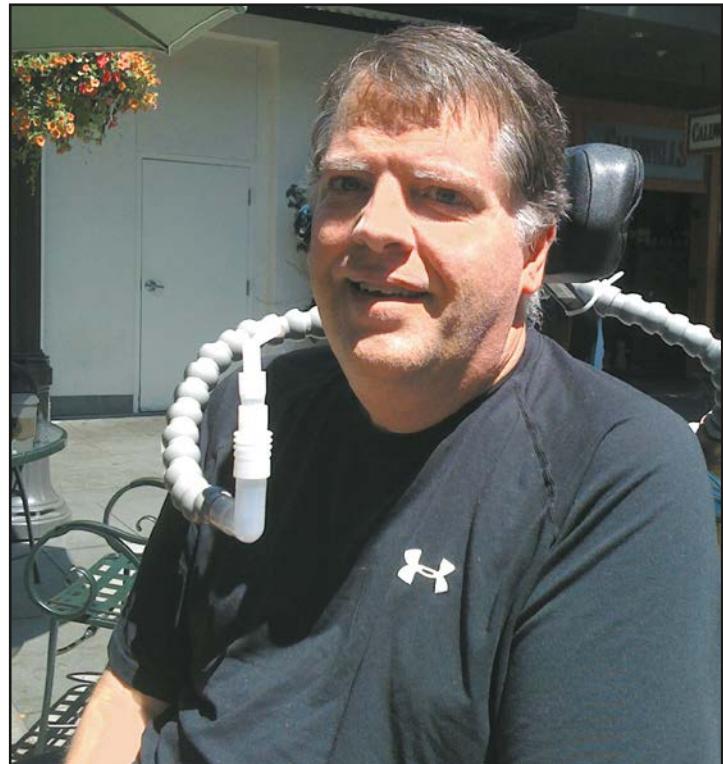
Dwyer rapidly lost the ability to breathe on his own and began using a Philips Resironics BiPAP in June 2007. But it wasn’t enough. Pat’s wife Jenny did an Internet search for “ALS and diaphragm” and found the NeuRx Diaphragm Pacing System.

About the DPS

The NeuRx Diaphragm Pacing System (synapsebiomedical.com/als/neurx-als.shtml) stimulates the respiratory diaphragm with electrical signals, by way of implanted electrodes. The system is designed to supplement breathing efforts and help preserve diaphragm muscle function.

In development for the past several years, in September 2011 the device received “humanitarian use” approval from the U.S. Food and Drug Administration for treatment of hypoventilation (inadequate breathing) in ALS. For more on the type of approval granted by the FDA, see “Physicians Push for Proof of Diaphragm Pacer Benefit,” page 216.

The approval means physicians can now prescribe the device, manufactured by Synapse Biomedical in Cleveland,



“It may be hard to jump through all the hoops,” says Dwyer, “but for us, the pacer was worth it.”

for people with ALS. It also clears the way for insurance companies to cover part or all of the cost.

Although the DPS doesn’t slow or stop the progression of ALS, it may improve quality of life. That was what Pat and Jenny Dwyer were about to find out, as part of the clinical trial of the device.

Getting into the DPS clinical trial

In 2007, Jenny Dwyer called to inquire about an ongoing DPS clinical trial, and three months later she and Pat flew to the lead trial site in Cleveland. There, Pat underwent a series of tests, which he describes as “pretty easy,” to determine whether he met the criteria to participate in the study.

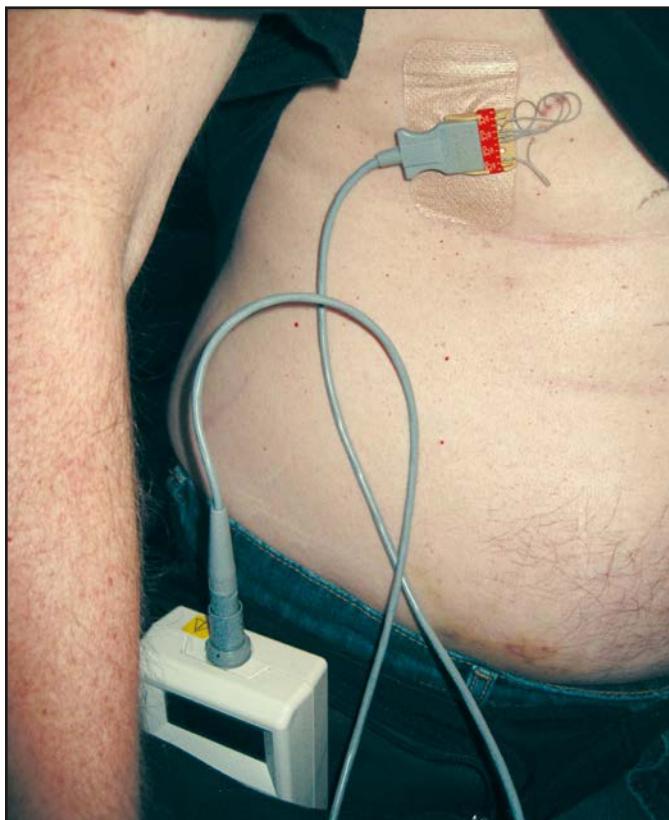
The entire battery of tests lasted between two and three hours and included:

- FVC, or forced vital capacity, which measures the maximum amount of air one can breathe in and out;
- blood carbon dioxide levels; and
- the “sniff test,” in which real-time observation of diaphragm movement is made using fluoroscopy (X-ray).

Because the results from the initial round of tests indicated that Pat had adequate preservation of his diaphragm and the phrenic nerves that stimulate it, he didn’t have to undergo separate phrenic nerve testing. (Phrenic nerve testing involves the use of various levels of electrical stimulation to different areas of the neck, and can be uncomfortable or even painful.)

Electrodes implanted via laparoscopic surgery

Pat met the criteria required to participate in the study. In December 2007, he and Jenny returned to Cleveland,



The external wires attach to the orange toggle, which is then attached to the pacer cord. The cord attaches to the pacer unit. Dwyer carries it in his pocket.

where he underwent minimally invasive laparoscopic surgery to have five electrodes implanted in his diaphragm. At the time he received the implants, Pat’s FVC had declined to 52 percent; an FVC lower than 50 percent would have prevented him from having the surgery.

In laparoscopic surgery, gas is pumped into the abdominal cavity in order to give the surgeon room to maneuver. Afterward, the gas has to be absorbed by the body and “that was a bit painful for Pat,” Jenny says. “He said he felt like it was all pushing up under his shoulders and trying to come out his armpits!”

When Jenny visited Pat in the recovery room, he was awake and talking, but pretty loopy from the anesthesia, she says. But all his numbers were good, and the pacer was all hooked up and ready to go.

Instructions for using the pacer

The following day Pat and Jenny spent approximately an hour learning how to turn the pacer control unit on and off, how to change the battery, and how to hook up the external wires to the pacer. The toggle with the external wires slides into the corresponding holes in the pacer cord. It only fits one way, so there’s no chance of hooking things up incorrectly.

They learned that getting the exposed wires wet when bathing is “no big deal.” While the area around the wires was healing, Pat kept it covered when showering. But now, Jenny says, “We just wash around it, and I’m careful when I dry it with a towel — just basically blot the wires dry.”

Flying home with the pacer was no big deal either, as the wires didn’t even set off airport security scanners.

Because Pat was a participant in the DPS clinical trial, he traveled back to Cleveland at three, six and 12 months post-surgery for monitoring. FVC measurements and the sniff test were repeated at all appointments.

A ‘shocking’ sensation

Pat first started “pacing” only four hours a day.

“You can feel the DPS working,” he says. “It feels like a low-voltage shock, but you soon get used to it and don’t really notice it anymore.”

Pat slowly built up until he was using the DPS 20 to 23 hours a day. As the pacing time increased, he experienced

a “sore-muscle” sensation in his abdomen but, as with the shocking sensation, he says he soon got used to it.

Pat says that when he began using the pacer, he felt less tired and lethargic at the end of the day, had more energy and didn’t need to nap as much as he had before.

But his FVC continued to decline, and he began using a sip-and-puff ventilator part time in September 2009.

“It was usually around three hours a day,” Jenny notes.

“Usually when he’d get home from work, he would ‘pump up’ with it before dinner.” Since then, Pat has progressed to using the ventilator most of the day, in addition to the DPS.

Prolonged independence and mobility

When Pat received the DPS, he still was able to walk, “but if he’d had to start using the sip-and-puff in 2007, he would have had to start using a wheelchair as well — probably a year earlier than needed,” says Jenny.

The wheelchair would have been required because the sip-and-puff vent weighs approximately 7 pounds and,



A “sip-and-puff” ventilator supplements Dwyer’s breathing efforts during the day.

says Jenny, “as much as I love my husband, I was not going to follow him around carrying his sip-and-puff!” The vent is now mounted on the back of Pat’s wheelchair.

The Dwyers believe the DPS helped Pat to continue running their business. (The couple owns two crab and salmon-on-fishery boats in Alaska). The extra time without the wheelchair also made easier a family vacation to Hawaii (where Pat was able to hang out in the pool and ocean), travel to horse shows, and time spent on the boats.

Another thing the pacer enabled Pat to do was sleep in a bed again. Before he received the DPS, he slept in a recliner chair because lying flat made it too difficult to breathe. Once he began pacing full time during the night, he was able to sleep in a bed with two pillows — “a benefit for both of us!” Jenny says.

The quality of his sleep is better too, Pat says, because the pacer and his BiPAP work together.

The here and now ... and the future

In December 2010, three years after he received the DPS, the pacer still was moving Pat’s diaphragm. “Not very much, and Pat had no movement on his own when the pacer was turned off, but there was still mechanical movement!” Jenny says.

Pat currently is dependent on his sip-and-puff ventilator during the day. He says the pacer works really well with both the sip-and-puff and his BiPAP and that there’s no sensation of the pacer “working against” either of the other two ventilation methods.

Although the couple has no way to tell whether the pacer is providing any benefit by keeping Pat’s diaphragm moving, Pat continues to use it.

“If he decides to stop using it,” Jenny says, “we would simply turn the pacer off and not hook it up to him.”

“While we know the pacer won’t stop the progression of the disease, we know that for us, it’s made our life with the disease a life better lived!”

Getting a DPS

The DPS is not easily obtainable, and not everyone will qualify for the device. ALS disease progression may be too advanced in some for the device to provide benefit.

Symptom progression also can make the required surgery too risky. Some physicians are calling for more evidence that the benefits of the device outweigh the surgical risks. Be sure to read “*Physicians Push for Proof of Diaphragm Pacer Benefit*” (below) for more information.

Currently there are only three DPS treatment centers in the United States: University Hospitals of Cleveland

(Ohio), Cedars-Sinai Medical Center in Los Angeles, and UT Southwestern Medical Center in Dallas. Another six centers provide referrals.

To view an interactive map containing locations and contact information, visit synapsebiomedical.com and in the “ALS Lou Gehrig’s” box click on “U.S. ALS Centers.”

Reprinted from the MDA/ALS Newsmagazine, January-March 2012

Physicians Push for Proof of Diaphragm Pacer Benefit

The U.S. Food and Drug Administration approved the NeuRx Diaphragm Pacing System as a humanitarian use device (HUD), a designation given to medical devices intended for use in rare diseases. For HUD approval, the FDA requires sufficient evidence the device does not pose “an unreasonable or significant risk of illness or injury, and that the probable benefit to health outweighs the risk of injury or illness from its use.”

FDA approval followed findings from a multicenter clinical trial that enrolled 106 patients with ALS and treated 86 for chronic hypoventilation at seven U.S. sites and one French site. Manufacturer Synapse Biomedical reported that study findings showed “the NeuRx DPS helps people with ALS live longer and sleep better than the current standard of care alone.”

Those who are interested in the device are strongly urged to discuss it with their ALS physician and find out as much as possible about the procedure before considering having it done. Although some physicians don’t think anyone with ALS qualifies for the DPS, because the surgery is too risky and benefit is unknown, others estimate that up to 25 percent will meet the FDA’s criteria to qualify for the device.

MDA/ALS center physicians polled about the DPS expressed some common themes.

More evidence is needed. Although the DPS has been shown to be safe in people with ALS, there remains a need for solid evidence that it actually provides benefit. Long-term follow-up must be done on those who participated in the Synapse trial, and additional well-controlled, large-scale clinical trials of the device are needed.



There are risks. Individuals must be aware that (as with any surgery, and especially when it involves someone with ALS) there are risks associated with the surgery to implant the pacer electrodes.

It is not a cure-all. The DPS does not prevent the need for BiPAP or other supplemental ventilation, nor does it stop or slow the progression of the disease.

“This is still a controversial procedure,” one ALS physician said. “It does provide some hope for ALS patients, but we must temper that enthusiasm with caution.

“While it is always optimal to have choices, it should be made very clear to patients that there is still no clear-cut choice when [considering] assisted respiration [options] in ALS.

“Patients must be provided with as much education and information as is available so they can make informed decisions on how they will approach their treatment.”

Appendix E

Empowering People with ALS to be ‘Research Ambassadors’

New training teaches individuals and families how to advocate for ALS research

by Nancy West

Fight smart, not just hard. That’s the motto of Richard Bedlack, a world-renowned ALS research warrior who directs the ALS Clinic at Duke University, Durham, N.C. “Everyone I’ve ever met who is affected by ALS wants to roll up their sleeves and do something about it,” he says.

Now individuals with ALS and their families have a new way to join the fight to find a cure for ALS. Last October, almost two dozen people attended the first annual ALS Clinical Research Learning Institute (CRLI), organized and led by Bedlack and Merit Cudkowicz, director of the MDA/ALS Center at Massachusetts General Hospital (Boston) and professor of neurology at Harvard Medical School. Cudkowicz also serves on MDA’s Medical Advisory Committee (MAC).

Learning research fundamentals

Designed to help those affected by ALS become “research ambassadors,” the CRLI featured faculty and staff from 10 ALS research institutions. This distinguished group outlined for participants the fundamentals of research, including trial design, ethics, informed consent, funding, regulatory affairs and enrollment barriers, as well as news about the ALS research pipeline and advocacy opportunities.

“We had open discussion of therapy development in ALS, what are the hopes, the challenges, and how we can work together to efficiently develop new treatments for people with ALS today,” says Cudkowicz.

A segment of the CRLI was devoted to understanding scholarly ALS research papers. Working in small groups, participants learned to critique a study. People with ALS want to keep up with the latest research articles “as much as we investigators do,” says Bedlack, but they don’t always have the background and training that allows for critical appraisal and interpretation.

“We want to teach them to find the hypothesis, dissect the methods used to investigate it, and be able to accurately assess the results and conclusions,” he says. “With this knowledge, our new research ambassadors are better prepared to go out and make contacts to help optimize ALS research in many ways, whether lobbying in Washington, speaking at support groups, or educating others with ALS about enrollment in clinical trials through blogs and online social networks.”

Addressing low enrollment

The idea for the CRLI grew from the discovery that only a small number of patients in ALS clinics have been enrolling in clinical research studies. “Yet, the only way to develop better therapies is through research,” observes Bedlack.

A study conducted by Bedlack and colleagues found that two of the biggest barriers to research enrollment are lack of awareness of research opportunities and misconceptions about the research process. Low enrollment in studies is a huge detriment to research because, as a result, trials take longer, cost more, and the results may not be generalized to the majority of people with ALS.

It’s very common for individuals with ALS to turn down participation in a clinical trial because they don’t want to be “guinea pigs,” says Bedlack.

Such ethical concerns were addressed at the CLRI training by James Russell, director of the neurology residency program at the Lahey Clinic, who discussed the protections now afforded to patients who enroll in research.

In settings such as ALS support groups, “We are counting on our new research ambassadors to help dispel the fears that patients have about research,” Bedlack continues. “By explaining the research process and discussing misconceptions, we can help them argue against those and other issues that are slowing our progress in finding a cure for ALS.”

Breaking down barriers with education and advocacy

Other barriers to optimal ALS research include funding limitations and government regulations, which Bedlack and Cudkowicz believe might be reduced through education and advocacy efforts.

“These efforts would be most effective if undertaken by large groups — not just scientists and clinicians, but also patients and caregivers who have the most at stake,” says Bedlack.

In developing the CRLI, Bedlack and Cudkowicz modeled it after a similar initiative that has been very successful in the Parkinson’s disease community. “We learned of their efforts to train patients and caregivers who are innately talented and charismatic — people who make others sit up and listen when they talk, and we sought out the same kind of people in the ALS community.” CRLI faculty members nominated patients and caregivers from within their practices to participate.

“So many people in the ALS community have so many diverse talents,” says MDA Senior Vice President of Advocacy Annie Kennedy. “Many are professionals who can no longer work because of their disease progression or because they are caregivers. But armed with knowledge from the CRLI, they can use their talent and ability to make powerful connections with people and advocate very effectively, in some cases without ever leaving home.”

Empowering people as advocates

One CRLI participant was Mary Murray, 66, of Braintree, Mass., who received an ALS diagnosis in January 2010.

“I feel empowered by the fact that the research community is looking to us to help spread the word about clinical trials and encourage participation,” she says. “ALS is often a fast-acting disease. So many patients get ‘too sick too soon’ to become active in the community. Those of us who have slower-developing symptoms must try to encourage new research to find new treatments for this disease. We also can encourage other patients with ALS to participate in clinical trials by helping them to understand what is involved.”

Murray has been participating in a clinical trial of dextropramipexole since April 2011.

One of the most powerful ways to advocate is to reach out to elected officials and tell your story. MDA provides a variety of advocacy opportunities, such as its yearly MDA Fly Out, which schedules visits to legislators when they return to their home districts from Washington D.C.; and its “Take 5” alerts about pending legislation of interest. (For more information about advocacy activities — both formal and informal — see “Use Your Talents to Advance ALS Research and Build Awareness,” page 219.)

“The power of patients is amazing,” says Bedlack, and Laura Tuttle, another CRLI participant, agrees. Tuttle, 49, of Raynham, Mass., learned in December 2009 that she has ALS. “It’s important for patients to give their point of view as well as clinicians. You can really have an impact when you tell your story,” she says.

Future opportunities

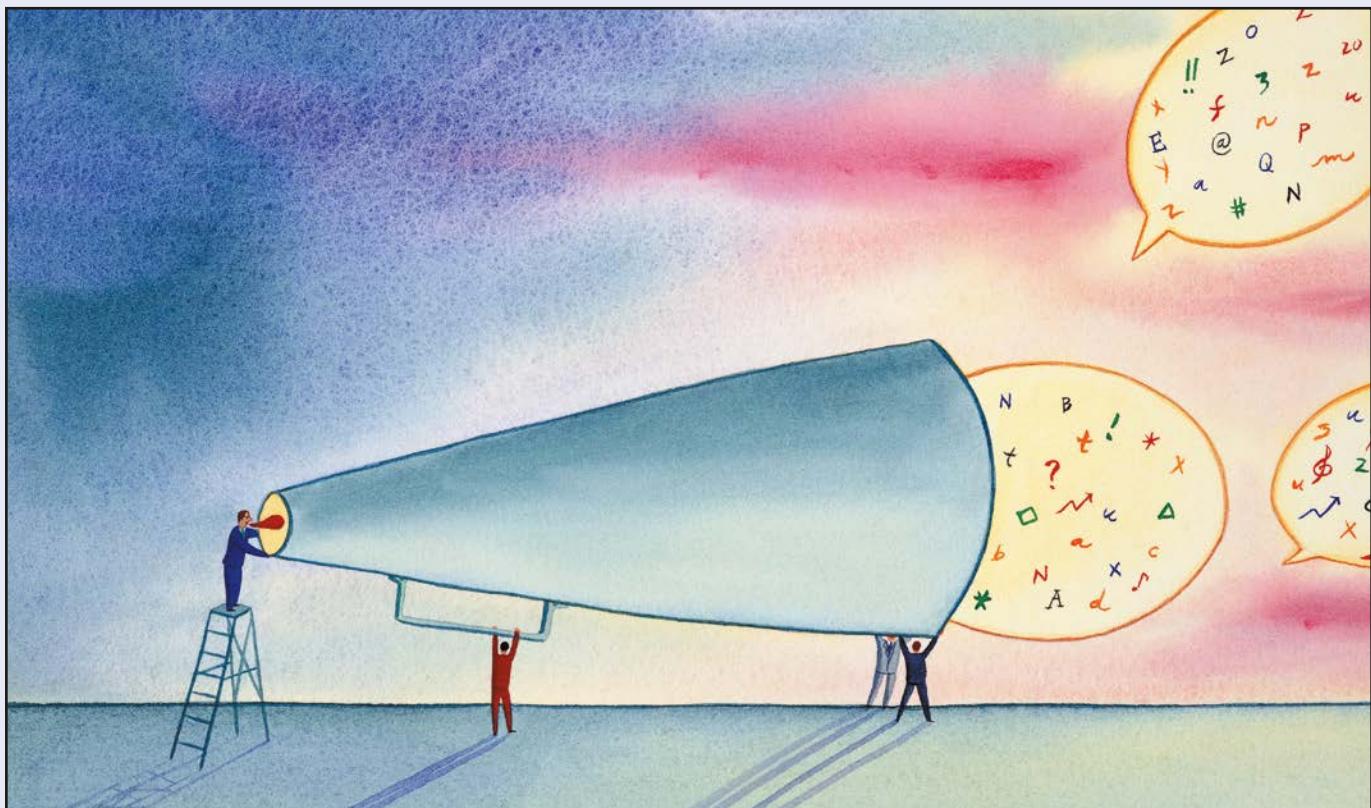
In the future, ALS research ambassadors may have opportunities to serve on federal committees such as the National Institutes of Health Advisory Councils that review research and grants, or U.S. Food and Drug Administration ad hoc committees convened for a drug review.

“When you sit on a committee as a layperson, you’re not expected to be a scientist, but you need to have an appreciation of the scientific process,” says Kennedy. “By providing that knowledge to our research ambassadors, the CRLI has helped to improve the likelihood that we will have people who meet the criteria to fill these committee openings as they become available.”

Bedlack also hopes to see CRLI participants blogging on the new blog space recently created on the Northeast ALS (NEALS) website.

“Attending a program like CRLI, targeting support groups, talking to Congress, participating in studies — to me, that’s a combination of fighting hard and smart that will help us move research toward a cure for ALS faster,” emphasizes Bedlack.

“The more information we each have about the disease and the newest work being done, the better,” adds Murray. “We cannot afford to let the new and exciting momentum in research suffer from lack of support — either the support of a knowledgeable and active community or financial support.”



Use Your Talents to Advance ALS Research and Build Awareness

Individuals with ALS and their families are a dedicated and talented group! There are many ways to use these diverse talents and skills to advance ALS research and build awareness of the needs of the ALS community.

No matter what your professional background or available free time, there's a role for everyone within MDA's ALS advocacy community.

Get involved with MDA

Become an MDA Advocate to receive timely updates on policy issues that impact the ALS community and to become involved in current advocacy campaigns. It's not necessary to have an ALS diagnosis to be an MDA Advocate — it's open to all. Register as an MDA Advocate and ask your friends, neighbors, coworkers and family mem-

bers to do so too. Let's ensure that the ALS community's voice is heard loud and clear by policymakers!

Stay informed about current research and clinical trials via the online MDA/ALS Newsmagazine, MDA website and your MDA clinic. Sign up to receive monthly email summaries of online research news.

Participate in MDA support groups and local outreach events, and take advantage of opportunities to inform others about research and advocacy news. Contact your local office to learn more.

Help plan an MDA fundraising or social event. MDA is proud to be the world leader in ALS research and services, and grateful to the hundreds of thousands of Americans who contribute year-round, helping to raise the vital funds that make these programs possible. Contact your local

MDA office to learn more about getting involved.

MDA's National Task Force on Public Awareness is an advisory body composed of individuals with neuromuscular disease who are leaders in their communities and achievers in their professional fields. The group serves as consultants to MDA on issues of concern to people with disabilities. Please notify your local MDA Health Care Services Coordinator if you'd like to be considered to serve on MDA's National Task Force.

Get involved in federal policy issues

Federal Advisory Councils: Individuals with ALS and their family members can participate on federal advisory councils and ad-hoc committees as consumer representatives at the National Institutes of Health, the Food and Drug Administration, and other federal agencies.

National ALS Registry: A working group of scientific and clinical leaders in the field of ALS, representatives from MDA and the ALS Association, and members of the ALS community serve as advisers to the Centers for Disease Control's oversight of the National ALS Registry. Input from the ALS community is the critical component in this collaboration.

Speak up: Opportunities for ALS advocacy can be found through letters to the editor; letters, calls and emails to your legislators; blogs; Facebook and other social media; and online communities.

Find your niche

The strength of the MDA ALS community depends on educated and empowered advocates. There's a critical role for everyone in the fight against ALS!

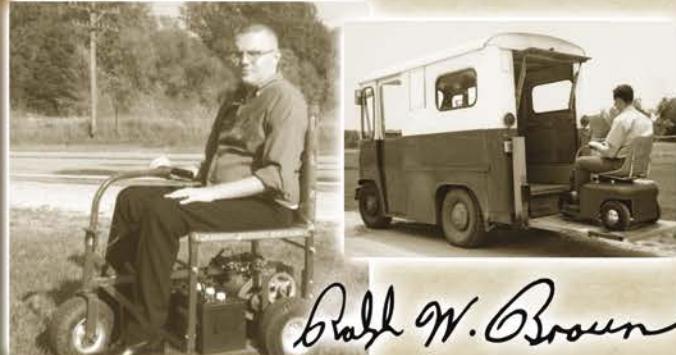
To learn more, contact MDA's Advocacy office at advocacy@mdausa.org, or visit mda.org/advocacy or call your local MDA office at (800) 572-1717.

Excerpted from the MDA/ALS Newsmagazine, January-March 2012



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