

A Manual For People *Living with* ALS

Third Edition

ALS SOCIETY OF CANADA

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AMYOTROPHIC LATERAL SCLEROSIS
SOCIETY OF CANADA

SOCIÉTÉ CANADIENNE DE
LA SCLÉROSE LATÉRALE AMYOTROPHIQUE

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MANUAL PREFACE

The purpose of this Manual is to provide information and helpful hints to individuals and families recently diagnosed and living with ALS. Readers are encouraged to use this manual as a tool to keep notes, organize personal information, and stimulate discussion between themselves, their family members, and their healthcare providers.

The content of the Manual represents an overview of ALS, coping tips and tools, where to go for help and support, and issues people diagnosed with ALS should begin thinking about early, such as assistive equipment that might be needed and legal and financial concerns for the future. In the area of clinical disease management, there is information about the healthcare professionals who make up the ALS care team, an overview of symptoms and functional changes associated with ALS, and specific information on a variety of disease management topics such as mobility, breathing, nutrition, oral care, communication, and end-of-life advance care planning.

Information about some aspects of ALS changes from time to time and understandably people want to be kept up-to-date with the latest information. However, sometimes news, particularly in the area of research and clinical trials, changes faster than the Society can update the manuals. To stay informed, the Society web site (www.als.ca), local ALS Societies, and ALS clinical specialists should be checked with from time to time for new information.

The Manual will be updated periodically, so readers with suggestions for updating or changing content, or altering the format to be as user-friendly as possible, are asked to complete and return the "Reader Feedback and Evaluation" form to share their insights.

It is our intent that reading this Manual will contribute to one's strength and encouragement. Strength from better understanding the challenges to be faced and encouragement from knowing that many others with ALS have found that even as the disease take its physical toll, their spirits have remained strong, surrounded by the love of their families and those who care for them.

ACKNOWLEDGEMENT

Previous editions of this Manual have been used as a source of information and support by many people across this country living with ALS, including those with the disease, their families, and caregivers.

This was especially true for its original author Bob Macdonald.

Bob had a unique exposure to ALS through his grandmother's, his father's, and his own experience living with the disease. When he was diagnosed with familial ALS, he plunged full force into the preparation of the first version of this resource manual.

His search for more information on the disease to expand on what he already knew led him around the world through his computer. The fruits of his search are evident here.

The 2005 edition of the Manual represents the third edition, which includes expanded and new information in an improved format.

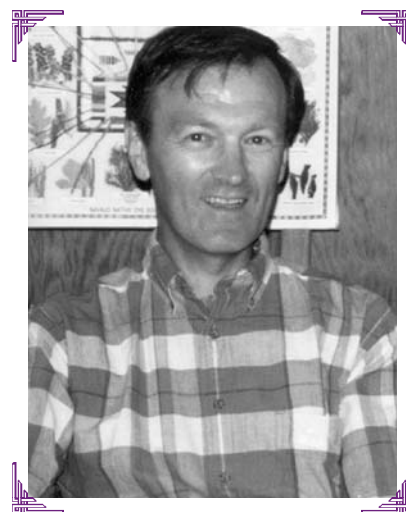
The ALS Society of Canada wishes to acknowledge the efforts of:

- The healthcare professionals with expertise in various areas of ALS management who were recruited to be contributing editors for their valuable input to ensure content quality.
- Members of the Services and Education Resources Committee (SERC) of the ALS Society of Canada for their direction and assistance in this endeavour.

And last, but certainly not least,

- The individuals and families living with ALS who gave of their time to pretest the Manual and provide their seal of approval.

This manual is dedicated to the memory of all those who have lived with ALS, but in particular to Bob Macdonald who held a vision for the original edition and inspired its continuation.



Bob Macdonald

(Photo and permission to use provided by Sandra Macdonald)

HOW TO USE THE MANUAL

ALS is not an easy disease to have. Reading about it won't be either. However, knowledge is a powerful resource. This Manual may be the first source of ALS information you are given. To get the most out of this resource tool, we suggest that you:

- Familiarize yourself with what is covered in the Manual so you know where to go when you need specific information-Do NOT feel as though you need to read it all at once
- Use the copy-ready personal assistance tools included at the back of Section 2 to help you and your family caregivers keep track of your personal details, needs, communication records, healthcare professional contacts, equipment records, medications, and appointments
- Share information in the Manual with friends and family who are interested in knowing more about ALS
- Consider the Manual a complement or additional resource to your primary source of information: your healthcare team of professionals who have a special interest in helping you manage ALS
- Review the "Resource Section" early to see what additional educational and support resources are available to learn more about ALS and specific aspects of coping with ALS
- Use information in the Manual to prompt discussions with your healthcare professionals about monitoring your progress and symptom management decisions
- Use the pocket page at the end of the Manual to store additional Fact Sheets and Research Updates as you collect them
- Use the binder cover pockets to store and keep together other materials you get from the ALS Society and elsewhere over time

The information for this publication has come from sources which we deem reliable and is submitted for general information purposes only. It is not intended to replace personalized medical assessment and management of ALS.

The ALS Society of Canada disclaims any liability for the accuracy thereof, and does not intend to disseminate either medical or legal advice. Throughout this publication, people with ALS are advised to consult with healthcare and legal professionals for medical and legal advice, respectively.

A MESSAGE OF HOPE

There is no doubt that living with ALS is challenging. However, with scientific technology speeding ahead faster and faster, there is more hope now than ever before that better treatments for ALS will be found, and that one day there will be a cure. But for today, know that there are organizations, health professionals, and others living with ALS who can help support you. As you live with ALS always remember:

- ALS may rob you of your physical body, but it does not rob you of your soul.
- You live in a society that emphasizes patient autonomy and you will be able to maintain yours.
- While embarking on a difficult journey, know you are not going it alone. Families and friends can become closer than ever, and you will discover new friends who understand.
- Other people with ALS are available and willing to share in the ups and downs of the journey.
- There are many dimensions to managing ALS and many professionals available to help you with them.
- The resources currently available to patients are without precedent.

- You continue to be a valued member of society who can contribute to your family and your friends through the human values in which you believe.
- You continue to play a role in educating your children and providing support to your family.
- Due to the intense interest in people with ALS, the options available to you exceed those that were available to previous generations and are expected to increase.
- While no one has chosen this path, most have traversed it with courage and with dignity.



DESCRIPTION OF ALS

Amyotrophic Lateral Sclerosis:

- A** = absence of
- myo** = muscle
- trophic** = nourishment
- Lateral** = side (of spine)
- Sclerosis** = hardening

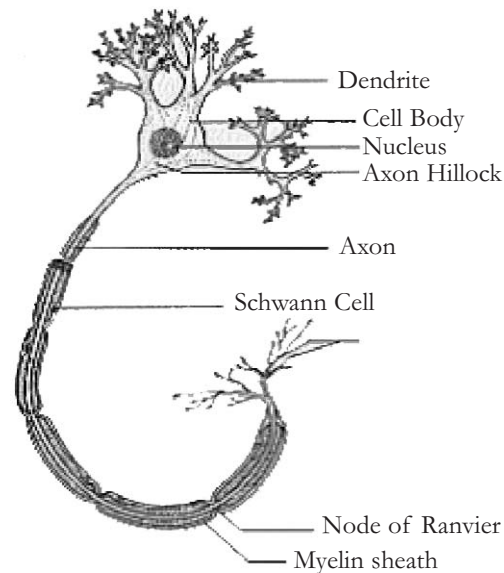
Dr. Jean-Martin Charcot, a French pathologist who founded the field of neurology, published the first full account of the symptoms of ALS in 1874. ALS is also known as Charcot's Disease, Lou Gehrig's Disease, and the most common form of Motor Neuron Disease (MND). In Canada, two to three people with ALS die each day.

Other types of MND you may know, although less common than ALS, are primary lateral sclerosis (PLS) and Kennedy's Disease (KD). PLS is different from ALS in that it does not involve muscle wasting, may progress over decades, and the expected lifespan may be normal. Kennedy's Disease, a form of adult-onset spinal muscular atrophy (SMA), is different from ALS in that it is a chromosomal X-linked recessive disease. Therefore only males are diagnosed. It is most often diagnosed between ages 20-40 years, progresses very slowly, and the expected lifespan is often normal.

Some people diagnosed with PLS at first may end up getting an ALS diagnosis after a few years if muscle wasting symptoms also occur. Sometimes people with Kennedy's Disease are misdiagnosed with ALS initially. An actual diagnosis of Kennedy's Disease is possible through DNA testing <http://www.kennedysdisease.org>.

ALS destroys motor neurons which are an important link in the nervous system. It is through motor neurons that the brain sends messages to the voluntary muscles throughout the body (muscles whose movement you can control as opposed to those you cannot, like the heart). Leg and foot muscles are controlled by motor neurons in the lower spinal cord. Arm, hand and finger muscles are controlled by motor neurons in the upper spinal cord. Speaking, swallowing and chewing are controlled by motor neurons in the brain stem. Respiratory muscles are controlled by motor neurons in the upper and thoracic levels (mid section) of the spinal cord.

The diagram below shows you what a motor neuron consists of:



ALS does not affect the five senses of sight, hearing, taste, smell and touch, nor does it normally affect the eye muscles, heart, bladder, bowel, or sexual muscles. There is no possibility that ALS is contagious.

ALS strikes about six to eight people per 100,000. With an estimated Canadian population of 32 million, approximately 2,200 people in Canada currently have ALS. In any given year, about two new cases of ALS per 100,000 people will be diagnosed. The incidence of ALS (new cases) increases with age (Strong and Shaw, 2003). ALS most often occurs between the ages of 40 and 70, but it can also occur in older and younger adults, and rarely in teenagers (Mitsumoto and Munsat, 2001).

TYPES OF ALS

The most common form of ALS is called **Sporadic ALS**. About 5-10% of cases are the inherited variety called **Familial ALS**. Until recently, an abnormally high incidence of ALS was observed in the Western Pacific (Guam, Kii Peninsula of Japan, Papua New Guinea). As a result of extensive research into Guamanian ALS, it is now thought that these high incidence rates were due to specific dietary toxins (for example, aluminum and cycasin). With the Westernization of this region, and the reduced dependency on traditional foodstuffs, major declines in the incidence of ALS in this region have occurred.

SYMPTOMS, SIGNS AND DIAGNOSIS

Symptoms are what you experience, or feel whereas **signs are what you can see or measure**. The early symptoms of ALS may seem vague. They can include tripping, dropping things, slurred or "thick" speech, and muscle cramping, weakening, and twitching. Some people with these early symptoms may tend to assume that they are normal signs of aging. As the disease progresses, the muscles of the trunk of the body are affected. Weakness of the breathing muscles develops slowly over months or years.

For some people, the muscles for speaking, swallowing or breathing are the first to be affected. This is known as Bulbar ALS. The term "bulbar" refers to the motor neurons located in the bulb region of the brain stem, that control the muscles used for chewing, swallowing, and speaking. ALS symptoms, and the order in which they occur, vary from one person to another.

The rate of muscle loss can vary significantly from person to person, with some patients having long periods with very slow degeneration. Although the average life expectancy from diagnosis is between three and five years, 20% live more than five years, and 10% live more than 10 years. Being a progressive disease, ALS may spread throughout the body over time, and at some point in this process, may involve the muscles required for breathing.

ALS can be difficult to diagnose in the early stages because symptoms may mimic other conditions. Because today there is not yet an available ALS-specific diagnostic test, other diseases and conditions

have to be ruled out first. However, there are clinical signs that can indicate wasting of motor neurons in either the upper or lower portion of the spine.

Doctors familiar with ALS usually see the following signs of lower and upper motor neuron degeneration:

Lower Motor Neuron Degeneration:

- muscle weakness and atrophy
- involuntary twitching of muscle fibres
- muscle cramps
- weakened reflexes
- flaccidity (decreased muscle tone)
- difficulty swallowing
- inability to articulate speech
- shortness of breath at rest

Upper Motor Neuron Degeneration:

- muscle stiffness, or rigidity
- emotional lability (decreased ability to control laughing or crying)
- increased or hyperactive reflexes

Some may assume that these are signs of normal aging. Over time, as muscles continue to weaken, and the weakening spreads throughout the body, it becomes more apparent that the cause is ALS.

In addition to a **physical examination**, people are often given an **electromyography (EMG) test, blood tests, a MRI (Magnetic Resonance Imaging), and other tests** to search for the presence of other diseases that can look like ALS.

Many doctors, including neurologists, do not have experience with recognizing and treating ALS. It is best to find out who in your province is familiar with ALS as soon as possible. Often patients are sent to ALS specialists to confirm a diagnosis and tell the patient they have ALS.

Telling someone and his or her family that they may have ALS requires a sensitive, understanding and compassionate manner. People have been diagnosed should be fully informed about the disease, treatments, current research and drug trials, and available support services, such as the ALS Society in their province.

See the **Resource Section** at the end of the Manual for books, videos, and web sites about ALS.

ALS. Three letters that change people's lives. FOREVER.

WHAT CAUSES ALS?

It has been well over a century since the first complete description of ALS by Dr. Jean-Martin Charcot. Today there is still no cure or effective treatment and the rate of ALS is on the rise. In spite of that painful reality, researchers are making rapid headway in understanding the complexity of the disease and developing future therapies. The accumulation of knowledge in the basic biology and potential mechanisms involved in ALS, coupled with impressive technological advancements, is accelerating the rate of progress in ALS research. It is commonly held that more advances have been made in the last ten years than in the last one hundred, and the sense of anticipation in the research community is stronger than ever before.

Once thought to be a single disease state, ALS is **now recognized to have multiple interacting causes, all sharing a common pathway leading to the destruction of the motor neurons.** By understanding the mechanisms that trigger this common pathway, we will ultimately understand ALS. Through such understanding, desperately needed therapeutic options will be developed. Already, new drug, gene, and stem cell therapies are in development. Because of the complex nature of ALS, it is thought that a combination of therapeutic strategies to attack the disease at all levels will ultimately provide the means to alter the course of the disease. In this section, we will highlight the potential factors and mechanisms that might play a role in ALS and their relevance to the development of such therapies.

Environmental Factors

Although ALS is age-dependent, and is on the increase as the average age of the population increases, the rate of increase is greater than would be predicted based on the aging population alone. This suggests the role of an environmental factor -- an idea supported by several examples in which clusters of ALS cases have occurred in a particular geographic area or environmental situation.

However, isolating specific environmental culprits has proven difficult so far. In the 1940s and '50s, a high incidence of ALS occurred in peoples in three regions in the western pacific: the Chamorros of the Mariana

Islands, the Auyu and Jakai of west New Guinea, and residents of the Kii Peninsula of southern Japan. An ALS-like disease also had an exceedingly high incidence on the island of Guam. The incidence in these areas seems to have decreased with Westernization, suggesting an environmental culprit, and dietary toxins have been suggested as a likely cause. A slightly increased rate of ALS also occurs in communities in southwestern Ontario where the water supply comes from regions where there is a major chemical industry.

Certain lifestyle factors such as diet, alcohol consumption, exercise, and smoking tobacco, and other environmental factors such as residence in rural areas, job-related exposure to certain toxins for ALS, physical trauma and pre-existing medical conditions have been studied as risk factors for ALS. To date, epidemiological research findings suggest that only smoking has shown a strong enough positive association to be considered a "probable," but not yet "established" risk factor (Shaw and Strong, 2003).

There is also an increased risk evident in those who have suffered a recent mechanical trauma. An association with electrical injury has even been postulated. It is hypothesized that viruses may also cause certain cases of ALS.

Although no single environmental agent has been shown to directly cause ALS, worldwide epidemiological studies consistently suggest environmental triggers are important enough to continue to study to better understand the relationships.

Because of the complex nature of ALS, it is thought that a combination of therapeutic strategies to attack the disease at all levels will ultimately provide the means to alter the course of the disease.

Genetic Factors and Inherited Variants of ALS

Approximately 90% of ALS cases are sporadic and do not show inheritance. However, **scientists assume that for many people who develop ALS, a genetic predisposition may interact with other factors such as environmental variables to produce the disease.**

A wide variety of naturally occurring genetic mutations could produce susceptibility to ALS. Recent research suggested genetic defects in dynein, a transporter protein in cells, could provide a common underlying mechanism of ALS and other forms of motor neuron disease.

In less than 10% of people with ALS, the disease is inherited. Seven genetic loci to date have been implicated in familial forms of ALS, each producing different features of disease in the families affected. Three of these genes have been precisely located and their corresponding protein identified (see table). The most common form of familial ALS is caused by the ALS1 gene, which codes for an abnormal form of the enzyme copper/zinc superoxide dismutase (SOD1) on chromosome 21. Researchers have shown that this mutation has not only a direct affect on the motor neurons, but on their neighbouring support cells as

well. ALS2 causes a form of juvenile ALS and maps to chromosome 2. ALS3 is linked to chromosome 18 and strikes in adulthood. The ALS4 gene on chromosome 9 is responsible for a rare form of familial ALS that affects boys during their teens and women in their thirties. The ALS5 gene also strikes mostly in adulthood and was mapped to chromosome 15 in 1998. ALS6 and 7 were discovered in 2003 and traced to chromosomes 16 and 20 respectively, and scientists are working to find their precise location.

Every newly identified and located ALS gene provides scientists a piece of the ALS puzzle and creates the opportunity to develop new mouse models and cell lines that simulate these genetic abnormalities to research the mechanisms that may occur in ALS. .

Even though inherited variants of ALS occur in a small percentage of ALS patients, these genetic models lend insight into the mechanisms of the disease as a whole.

Gene	Type	Chromosome	Year Found	Affected Protein	Reference
ALS1	dominant, adult onset	21	1993	Cu/Zn superoxide dismutase (SOD1)	Nature. 362:59-62
ALS2	recessive, juvenile onset	2	2001	GTPase regulator "alsin"	Nature Genetics. 29(2):166-73.
ALS3	dominant, adult onset	18	not yet		Am J Hum Genet. 2002 Jan;70(1):251-6.
ALS4	dominant, juvenile male onset, adult female onset	9	2004	DNA/RNA helicase	Am J Hum Genet. June; 74(6).-in press
ALS5	recessive, adult onset	15	not yet		Neurogenetics. 1998 Dec; 2(1):55-60.
ALS6	dominant, adult onset	16	not yet		Am. J. Hum. Genet., 73:397-403, 2003
ALS7	dominant, adult onset	20	not yet		Am. J. Hum. Genet., 73:397-403, 2003

Genetic testing of the SOD1 gene is clinically available. Current testing is only able to identify the known genetic loci. Even though some loci have been identified, the majority of cases of inherited ALS remain unexplained. There are research projects that seek blood samples from persons with familial ALS to further their work in identifying more genetic loci and better understand the disease process. Ask your neurologist for more information about these studies. Testing is appropriate in any patient with ALS who has another affected member or an incomplete family history. Molecular genetic testing could clarify mode of inheritance (i.e., autosomal dominant, autosomal recessive, or x-linked dominant determined by family history) and modify risk assessment for genetic counseling as well as perhaps indicating disease prognosis.

Genetic testing should always be accompanied by formal genetic counselling. Genetic counselling is the process of providing individuals and families with information on the nature, inheritance, and implications of genetic disorders to help them make informed medical and personal decisions. **Genetic counsellors** are specially trained professionals who use family history and genetic testing results to clarify genetic risk for other family members and help you weigh the pros and cons of testing.

Presymptomatic testing for SOD1 mutations for adults, who have no symptoms of ALS but do have a family history, is available but controversial. It is controversial because of incomplete **penetrance** (penetrance is a term used in genetics that describes the extent to which the properties controlled by a gene will be expressed); inability to predict age of onset, the lack of preventive measures, and to date there is no cure for ALS. For these reasons, getting tested and receiving a positive result (that is, the person does carry the gene) may cause undue stress and anxiety about when ALS symptoms will strike. **Testing of children** requested by parents requires sensitive and understanding counseling. A consensus exists that asymptomatic children should not be tested as it removes their choice later in life and increases the chance of social stigmatization, and could negatively impact educational and career decisions.

If you are thinking about genetic testing, ask your neurologist about where you can go for genetic counselling

and testing, or self-refer to a genetic counsellor. For more information, visit www.genetests.org

Free Radicals and Oxidative Stress

All cells generate toxic metabolites (think of these as the "exhaust" from your car and the consequences to yourself and your passengers should your car not be properly vented). In the cell, these "exhausts" ("free radicals") are a destructive form of oxygen that can also be used by the cell under normal circumstances to fight disease. However, an overproduction of free oxygen radicals can result in cell damage and death. As might be expected, several defenses against such a process exist. A major one is the superoxide dismutase enzyme (SOD-1) discussed above. Mutations in the SOD-1 gene could allow excessive free radical production, damaging the neuron. Canadian researchers have documented the existence of excessive levels of proteins damaged by free oxygen radicals within neurons in ALS, suggesting that either the neuron synthesizes excessive levels of free radicals, or that it is incapable of "venting" those that are normally produced.

Therapies geared to reducing oxidative stress are in development, including gene therapy and new pharmacotherapy. The properties of certain foods and vitamins are also studied for their ability to reduce free radicals. Vitamin E's antioxidant properties made it the prescribed therapy for people with ALS one hundred years ago and it still is today, although with little effect in hindering the course of the disease.

Immunological Factors

In recent years, researchers have considered how injured motor neurons might spur an immune response that could contribute to, and even perpetuate, a cascade of cell death in the nervous system. The immune cells called microglial cells and astrocytes can respond to neural injury in a way that can be either beneficial or harmful. This microglial response has been implicated as a trigger of programmed cell death (PCD), a mechanism that is useful on the small scale to clear away damage, but devastating on a large scale as it ripples through the nervous system killing motor neurons.

Neurotrophic Factors

These important factors for the growth and maintenance of motor neurons in humans have been shown to enhance motor neuron survival in mice with a variety of motor neuron disorders. Although it is not clear how deficiencies of neurotrophic factors may affect human motor neurons, several attempts have been made to determine whether neurotrophic factors can slow the rate of progression of ALS by first testing these agents in animal models of the disease.

Researchers are also investigating how the neurotrophic factors including brain derived neurotrophic factor (BDNF) and cytokine ciliary neurotrophic factor (CNTF) **interact with metal ions in cells, and how metal ions can thereby have extremely toxic effects in a cell depending on the neurotrophic factors present.** By developing drugs to manipulate the interactions of neurotrophic factors, researchers hope to learn about and gain some control over this potential aspect of ALS.

Gene therapies are also in development to promote the levels of beneficial neurotrophic factors. Recently, the gene for insulin-like growth factor 1 (IGF-1) was successfully delivered in ALS mice using a viral vector, with the successful result of prolonging the life of the mice.

Altered Protein and Neurofilament Metabolism

Advances in technology in the field of protein research have opened up new avenues into understanding the **protein mechanisms** involved in ALS. **Mass spectrometry**, which enables scientists to detect which proteins are present in an extremely small sample, has become very advanced. Mass spectrometry is also providing researchers with the tools to develop a desperately needed simple, **rapid test to diagnose ALS** by identifying proteins generated through the course of the disease. Powerful electron microscopy reveals the configuration and behaviour of proteins. Such technological advancements provide insights into how proteins fold and unfold and interact with their environment.

A signature feature of ALS is the accumulation of neurofilaments in the motor neurons. These key neuronal proteins are believed to be responsible for maintaining the normal neuronal structure and shape.

Studies making use of transgenic models to alter neurofilament expression reveal that abnormalities in the metabolism of neurofilaments, or the way in which neurofilaments interact with each other -- or with other proteins, could play a role in the development of ALS. Many types of cellular proteins and enzymes may play a role in ALS. Protein kinases are enzymes within the brain and spinal cord that regulate many cellular functions. Studies have revealed that abnormal levels of protein kinases exist in the nerve tissue of people who died of ALS. By comparing this tissue with mouse models, researchers are investigating how skewed levels of these important regulatory enzymes may trigger cell death in the motor neurons in the hope of finding a way counterbalance the protein kinase ratios to prevent cell death from triggering.

Proteosomes and protein chaperones are enzymes that shuttle proteins and chop them up into their basic components in order to clear away damaged proteins in the cell. Researchers are investigating these enzymes in mouse models and cell lines to understand their role in the pathogenesis of ALS and how they might be used therapeutically to hinder the disease.

By studying the proteins affected by genetic mutations in familial ALS, and the behaviour and interactions of the proteins involved in neurofilament aggregation in cells, **researchers are gaining a better understanding of the role of abnormal protein mechanisms in ALS.**

Glutamate Excitotoxicity

Abnormalities in the handling of excitatory amino acids by the nervous system, particularly glutamate, may be critical to the occurrence of ALS. Through damage to the normal "transporter" mechanisms by which glutamate is removed from the nervous system, excessive glutamate accumulates. When motor neurons receive glutamate at their receptors, there is an influx of calcium ions into the cell. The motor neurons may not be able to deal with the excessive levels of calcium flooding in, resulting in damage. Researchers are investigating ways to help the nervous system handle calcium and glutamate. Riluzole, the single drug currently available for the treatment of ALS, shows very modest results. Its action is not well understood but is thought to perhaps affect glutamate mechanisms.

ALS. Three letters that change people's lives. FOREVER.

IS ALS TREATABLE?

Clearly, there is a critical need to find treatments to effectively slow the progression of ALS, or cure it completely. However, with today's quickly advancing scientific technology, the odds of finding effective treatments for ALS are better than ever before.

Drug and Clinical Trials

Medications used to treat a disease are based on the results of laboratory and clinic-based research studies, or trials. Drugs are first developed in the laboratory using technological methods and animal model testing to see if the treatment has an effect on the disease process. Treatments that show promise then need to be studied on actual patients with the disease in a clinical setting to test for **drug delivery efficiency** (i.e., what is the best way to give the medicine?), **safety** (i.e., how well is it tolerated and at what dosage?), and **efficacy** (i.e., is the drug actually changing the expected course of the disease in patients receiving the drug as compared to others who are not and to what degree is there a difference?). **While not all clinical trials are successful at turning out a new, acceptable treatment, they nevertheless help to answer research questions and move research in a forward direction.**

Clinical trials are subject to stringent research protocols for the protection of human subjects. From time to time there may be an opportunity for you to participate in a trial. To learn more about the clinical trial process, refer to Fact Sheet #12: *Clinical Trials-A Primer* included at the back of this *Manual*. This fact sheet can also be downloaded from the ALS Society of Canada web site, www.als.ca. The Fact Sheet covers why trials are important, the four phases of trials, how trials work, who conducts them, safety issues, risks and benefits, informed consent, and questions to ask your doctor.

Ask your neurologist for up-to-date information on what clinical studies are currently being conducted to help us better understand how to treat ALS. A database of worldwide clinical trial information can be found through the World Federation of Neurology at www.wfnals.org.

Final Note on what is ALS?

In summary, ALS is a complex disorder in terms of how and why motor neurons are destroyed as well as how different individuals are affected. Given this complexity, ALS research can be daunting. However, scientists continue to make progress in better understanding the disease pathways. Likewise clinical researchers continue to conduct trials to gain more insight into effective therapies. With scientific technology moving ahead at a very rapid pace, there is more hope than ever in the fight against ALS.

From time to time there may be an opportunity for you to participate in a trial. To learn more about the clinical trial process, refer to Fact Sheet #12: *Clinical Trials-A Primer* included at the back of this *Manual*.

THE ALS DIAGNOSIS: NOW WHAT?

Many people with a serious disease will feel torn between keeping a positive attitude to keep illness away and knowing that at some time they have to accept they do have a serious disease. When the existence of disease is known, "fighting it" is another approach some people take. Others prefer to avoid a win-or-lose approach, and instead deal with things day by day.

An ALS diagnosis has the power to strengthen healthy family relationships, or shatter already weak ones. It can unpredictably bring out the best in some people and in others, awaken emotions they cannot manage. To help you cope and instill hope, the ALS Society is here for you and your family (**Section 3**). This section is intended to give you suggestions for personal coping, as well as tips for others around you who will also be going through changes with you. **Section 4**, sub-section *Approaching End-of-Life Issues and Advance Planning*, and **Section 6** *Legal and Financial Issues* also provide information that may help you and your family to cope with changes and planning ahead.

COPING STRATEGIES FOR THE PERSON WITH ALS

Social workers, psychologists, counsellors, and support groups can be very helpful. You can learn from those who are familiar with what you are going through, share understanding with others who have the same challenges as you, and learn how people with more advanced ALS are dealing with issues you might face in the future. The ALS Society, hospices, and other organizations provide these types of services to help provide social and emotional support throughout the course of the disease.

One's life with ALS can be looked upon as a series of progressive functional losses, but it can also be a unique opportunity for enrichment. The choice is yours. If you choose enrichment, it can take many forms. The choices are very personal.

You may decide to develop closer relationships with family and friends; make new friends with others who are sharing your experiences and with many other people who are involved with ALS; learn about computers and how to communicate with others through the

ALS. Three letters that change people's lives. FOREVER.

CHANGES...

Changes will take place for everyone. Relationships with family and friends, children and colleagues, will undergo a transition period. Close friends and relatives, including children, should be told the truth about your ALS. For most people, not knowing what is happening is worse than knowing the truth. Children in particular imagine frightening things if they can see that someone is sick and no one talks about it in front of them. Knowing what is happening also makes it easier for people to offer support and to help. Don't be afraid to ask for help when it is not offered; people often don't know what to say or what would be helpful. Most people, especially relatives, are glad to be of assistance.

Telling others about your situation often means that you will discover who your true friends are. Some people find it very hard to be around sickness at any time, and others simply do not want to commit themselves to helping in a serious situation. Expect that some people may break off relationships. Although this is hurtful, it should not be taken personally. Most people will try to help and be supportive.

- ALS: Strategies for Living
ALS Society of British Columbia, 1993

Internet and other similar services; learn to appreciate other things in the world around you, things that you previously took for granted; take time to read and listen to music and learn more about spiritual awareness. The list can go on and on. It won't likely include everything you've always wanted to do, but it can include enough to give you a full and satisfying life. It all depends on your attitude and imagination.

Hope, faith, love, and a strong will to live offer no promise of immortality, only proof of our uniqueness as human beings, and the chance to experience full growth, even under the grimmest of circumstances. The clock provides only a technical measure of how long we live. Far more real than the ticking of time is the way we open up the minutes and invest them with meaning.

Meeting the Challenge

Acceptance of ALS does not mean giving up. It should be the first step in making the most of your life with ALS. There is much that can be done to help you live a productive and enjoyable life. Be positive but do not trivialize the situation. It isn't helpful to pretend that everything will be fine or that ALS is not a very serious condition.

On the other hand, there is no need to dwell on the negative aspects of the disease. Twenty percent of people with ALS live more than five years and nearly 10% live ten years or more. You could be one of those people. And thankfully, neurological research is moving steadily ahead, so no one knows when a breakthrough may occur...it could be sooner than later. These facts give hope and hope is a crucial part of life. Achieving a balance between hope and realism is difficult at times, but something we all must learn to do.

If you are a person with ALS who has decided to take charge of your condition, **here are a few practical things you should consider doing without delay.**

1. Contact your family doctor

When doctors find out that one of their patients has ALS, some might say there is nothing they can do to help. In fact, they probably can help but they don't know it! If your doctor hasn't already done so, request that he or she refer you to the nearest ALS Clinic or ALS Rehab team. Your doctor will also be able to help you in other ways in the future, such as signing your application for a CPP disability pension, and the Canadian Revenue Agency (CRA) Disability Tax Credit Certificate for your annual tax deduction, or by approving your application for home care.

You should also suggest that your doctor access the resources from the ALS Society by requesting a copy of this manual or other information the ALS Society makes available. Also encourage your doctor to rely on specialists and therapists associated with an ALS clinic or centre for consultations and advice. You also need to discuss with your family doctor his or her availability to do home visits as ALS progresses.

2. Register with the ALS Society

The ALS Society provides various services to assist people with ALS. Let your local provincial ALS Society know that you are a person with ALS in their area, and that you would like to register with them and receive any helpful information they provide.

3. Apply for a Disabled Parking Permit

Your local city or provincial government will issue a disabled parking permit to people with ALS. Get the application through your local ALS Society or ALS Clinic.

4. Obtain a Medic Alert Bracelet

A Medic Alert bracelet can be useful to tell others about your condition, in the event that you are unable to speak. This may be particularly useful for those with Bulbar symptoms who are unable to speak or have slurred speech. For further information and an order form call the Canadian Medic Alert Foundation at 1-800-668-1507.

5. Contact your auto insurance provider

If you don't report your disability to your insurance company, your auto insurance coverage may not be valid. Driving sometimes requires fast foot and hand reactions. Enquire about a driving test service in your community that will certify your current abilities for insurance purposes.

TO DO LIST. . .

1. Contact your family doctor
2. Register with the ALS Society
3. Apply for a Disabled Parking Permit
4. Obtain a Medic Alert Bracelet
5. Contact your auto insurance provider

Tools for Staying on Track

You can help yourself cope by staying organized and keeping track of your changing needs, who you have been in contact with and for what, appointments you have, medications you take, etc. To help you stay on track, several **personal assistance tools** have been included for you at the end of this section. Each one is a copy-ready form. Do not write on the original, but use for photocopying. Keep your working forms in a file at home for easy access by you and your family caregivers. Some people find dedicating an accordion file case specifically for your ALS care management files helpful.

The following tools are included in this section:

- **Personal Details**

This lists your contact information, date of birth, living arrangements, caregiver information, and type of ALS. This is helpful to share with healthcare professionals you are coming into contact for the first time as repeating this may become tedious.

- **"About Me"**

On this page you can describe how ALS is affecting you and your current individual needs. This may be helpful for nursing staff if you are hospitalized, or for people who come into your home to care for you.

- **Communication Record**

This is a handy tool to document what healthcare professionals have said to you when they come to visit you, or when you go to visit them. If you prefer that they document what they have said or what they say they will arrange for you, ask them to use your form. It is also a way for them to see what other types of visits you have had or what referrals have been made for you so that efforts are not being duplicated.

- **Professional Contact Sheet and Cardholder**

You may choose to use both or one of these tools as a mechanism to keep your healthcare professional contacts organized. Retain the name and contact information for all professionals you see either in the clinic, through home care, community care centres, hospitals, etc.

- **Equipment Record**

Over time, you will likely find the need for various pieces of assistive equipment. Realistically, you may acquire different items from various sources. This form provides a place to document by date each item received, whom you need to contact about that item, and their phone number.

- **Medication List**

This form allows you to keep track of prescribed and over the counter medicines you are taking. You may find it useful to take this form with you when you go to the doctor to jot down information about how to take the medicine and if there are any side effects or alternatives.

- **Appointments Log**

You may already have your own system of keeping track of appointments in a day planner or electronic device. If not, you may want to use this log to jot down specifics regarding appointments you have made relative to your ALS care.

These tools were adapted from *Your Personal Guide to Motor Neurone Disease*, Motor Neurone Disease Association, Northampton, UK
www.mndassociation.org

COPING STRATEGIES FOR FAMILY MEMBERS

It is important to be aware that members of your family and your friends are experiencing emotional reactions to your disease. They may feel guilty

that you have ALS

Tips and Advice:

and they are healthy. They may also be short-tempered because of the extra daily responsibilities that they face -

banking, raising children with less help from you, more chores - all this in addition to the care they give you. They may feel that it is not fair and then feel guilty about reacting this way.

See the following Fact Sheets that have been included at the end of the Manual, and the *Resource Section* for helpful books and web sites.

#8-First Steps for Families:

For families of those recently diagnosed with ALS

#9-Ways to Help:

Assisting Families living With ALS

arrangements.

The needs of a caregiver tend to take a back seat to the needs of the sick person. It is hard to have a serious illness, but it is also difficult to care for someone with one. Most people, sick or well, feel a combination of some or all of the following emotions at different times when dealing with illness. Most importantly, no one should feel guilty about any of these feelings. They are perfectly normal and to be expected.

Keeping the lines of communication open is the best way to work out these feelings. Talk openly to your family about how you feel. Encourage them to share their feelings with you. If this is hard to do, it may be useful to talk with a social worker or other member of your ALS team. He or she may be able to help you and your family to solve communication problems. It is most important that you have someone to talk to who will not be upset by what you are saying. The person you need may be one of your healthcare professionals, a friend or a family member.

Members of your immediate family will likely become involved in your daily care and assistance. Most people with ALS remain in the home for as long as possible, and the demands upon family members can be great. Your primary "informal" caregivers, such as your spouse, partner or grown children, may find that your care, especially in the later stages of the disease, is taking up most of their lives. It is natural for them to want to give all that they can to help you, but there should be limits to self-sacrifice.

Caregivers must continue to have lives of their own and take care of themselves. Spending time in the company of friends or family who are not sick, in the pursuit of hobbies or activities in addition to care giving, or quiet time alone are important refreshers. These periods of respite allow caregivers to recover from the stresses of care giving and make them more effective, balanced helpers. Do not hesitate to ask

other family members to fill in for your primary caregiver while he or she takes regular breaks or, if possible, arrange for paid help. A social worker or ALS Society representative may be able to help you identify respite services and any available funding

Commonly experienced emotions in both the initial and longer terms are:

Initial emotions:

- Curiosity
- Love
- Closeness and openness to others
- Hope
- Disbelief
- Loss
- Anticipatory grief
- Guilt
- Erosion of trust
- Denial
- Responsibility

Long-term emotions:

- Persistence
- Hope
- Love
- Appreciation of the value of life and of others
- Sadness
- Guilt
- Loneliness
- Jealousy
- Annoyance
- Feeling trapped
- Feeling overwhelmed

Remember, everyone has the right to:

- put themselves first
- sometimes make mistakes
- have their own opinion and convictions
- change their mind or decide on a different course of action
- protest unfair treatment or criticism

- *List excerpted from ALS: Strategies for Living, ALS Society of British Columbia, 1993*

HELPING CHILDREN COPE

Although ALS is not primarily a disease of the young, it sometimes happens that the family of the person with ALS includes young children. It is easy to forget that they need to know how the disease will affect you

ALS: It's all in the family...

...Talk as openly as possible around children, as secrets make everyone uncomfortable.

...Encourage children to talk about how they feel, to cry if they want to, and to talk about the situation to anyone with whom they feel comfortable. Children may hide their feelings at times, but that does not mean that they do not care, or are not feeling anything.

...Encourage children to help when they express the desire to do so. Being useful will make them feel involved and valuable. They do not have to help with medical care: Doing more chores at home or just reading and spending time with their relative with ALS are all possibilities.

...Make sure life goes on as normally as possible for children (and don't be afraid to ask others to help you make this happen) like keeping them in activities they have always done, spending time with friends, going out as a family.

...Allow children to be involved in planning family activities that will include the relative with ALS. This helps them feel useful and better understand lifestyle changes that must be made.

See the following Fact Sheets included at the end of the Manual:

#4-Caregiver Stress:

10 Signs of Caregiver Stress

#5-Reducing Stress:

Reducing caregiver stress, finding ways to feel better

as a beloved parent or grandparent, and the whole family. It is important that your family members and counsellors helping your family take time to work through the problems children may have in coming to terms with ALS in their family. **Two resources for children** available through the ALS Society include *A Booklet for Young People*, an ALS Canada publication, and *Grandpa, What is ALS?*, produced by the ALS Society of Alberta. Contact your provincial ALS Society to find out more about how you can acquire these books. A third resource, *Someone You Know Has ALS*, is available through the McMaster Medical Centre (905-521-22100 x76870).

The feelings of adults and children in upsetting situations are quite similar. Anger, helplessness, fear, hope, and despair are commonly experienced. Children, however, have fewer ways of expressing their emotions and dealing with them. Younger children tend not to express themselves in words and will act out their feelings. Older children can find it hard to talk about how they feel, and teenagers often do not have friends with whom they can talk about serious situations.

For children of all ages, feelings can be expressed through emotional and behavioural changes. Children may have questions that they may be afraid to ask because everyone is so upset, and they do not know how to find things out by themselves. Although many people feel that children should be protected from sickness, this does not help the child. The child will know that something is wrong and will get upset anyway.

Talking to Children

When dealing with children experiencing sickness and big changes in their families and lives, it is necessary to explain everything as fully as possible. **Because adults tend not to explain to children what is happening, children often blame themselves for the sadness in the house and feel very guilty.** They may want to know what exactly it is that the sick person has, whether they can catch it too, what will happen to the sick person, and whether that person will die. These questions should be answered as positively as possible. For example, instead of telling the child that a person is very sick or dying, something like: *"Well, they are not doing very well right now, but the doctors say that there are still things we can do. We can....."* gives hope without being dishonest.

Children should know that they can ask questions and that someone will try to answer them. If their parents do not feel able to talk to them, then a relative, friend, or doctor should make sure that the child knows that someone is always available to talk, not only about the illness but also about anything that is worrying the child.

A major concern of children is often what will happen if the sick person becomes very sick or dies. They may want to know if they will have to move, who will look after them, if they will have to give up their favourite things, change schools, and so on. They may be afraid to ask about these things, so they should be reassured and told about any planned changes before a crisis interrupts their normal routine.

Very young children may be upset by what is going on, but will probably not be able to understand your explanation. Giving them lots of hugs, love and attention helps until they are old enough to understand the situation in more detail. **Whatever transpires, do your best to keep the children involved and feel secure.** Do not hesitate to turn to members of your healthcare team or the ALS Society for advice or help when you need it. The web site www.alsindependence.com has excellent information on how to explain ALS to children of various ages.

Long-term ways to help children cope...

...**Pay adequate attention to children of parents with ALS.** This may mean asking a grandparent or other family member or close friend to help you with this.

...**Explain to children that a change in appearance does not mean the sick person's feelings toward them have changed.** Protect children from the emotional swings of the sick person, if mood changes are occurring.

...**Allow children respite, too.** Like caregivers, they need a break to enjoy themselves.

...**Continue to encourage children to express their feelings.**

Final Note on Coping...

ALS, like many chronic diseases affect the person with the diagnosis and those around them. Each person needs support and resources to learn, accept, and cope with the changes. The Educational Resource at the end of the Manual includes many books, videos, and web sites that may be helpful to you. Because ALS is a progressive condition, change is on-going. Therefore, tools to help you cope may be useful at various points in time when you are living with ALS.

Personal Details

Name:	Date of Birth (DOB):	Date of ALS Dx:
Contact Information:	Lives Alone:	<input type="checkbox"/> Yes <input type="checkbox"/> No
<u>Address:</u>	Lives with:	<u>Relationship:</u>
	<u>Name/s:</u>	
Phone number:		
Cell number:	Dependents:	<u>Relationship:</u>
E-mail:	<u>Name/s:</u>	
Health ID #:		
Private Insurance Plan/ID #:		

Informal Caregivers (family, friends, neighbours, etc.)	Type of ALS:
<u>Name/s:</u>	<input type="checkbox"/> Familial ALS
<u>Address:</u>	<input type="checkbox"/> Sporadic ALS
<u>Phone:/E-mail</u>	<input type="checkbox"/> Don't know
	Form of ALS:
	<input type="checkbox"/> Limb onset
	<input type="checkbox"/> Bulbar Onset
	<input type="checkbox"/> Don't know

About Me...

You can use this page to let people know about your individual needs and how ALS currently affects you. This may be particularly helpful for nursing staff if you have a stay in hospital or a respite care facility, or require homecare. For example, you may wish to include what foods you can eat, signals you use to communicate, etc.

Professional Contacts

Professional Contacts					
Title	Name	Address	Phone	Fax	E-mail
Family Doctor					
Neurologist					
Neuromuscular Nurse					
Occupational Therapist					
Physical Therapist					
Speech & Language Therapist					
Dietitian					
Social Worker					
Community Care Coordinator					

ALS SOCIETY OF CANADA

The ALS Society of Canada was founded in 1977 as a national non-profit, charitable organization dedicated solely to providing a variety of services to people with ALS and their families. The Society has affiliated Societies in every province in Canada and remains the only organization in Canada dedicated solely to those affected by ALS.

In effect, the Society is mandated to help you today through *information* and *support*, as well as tomorrow through funding *research* that will change the course of ALS and ultimately put an end to it.

Providing Information

The ALS Society of Canada provides **comprehensive, accurate, and timely information** about ALS to those with ALS, those affected by ALS, the research and healthcare communities, the media, our donors, and volunteers.

Our Mission:

We communicate through several publications such as our newsletter, *Coast to Coast*, our research publication, *The Northern Neuron*, brochures, fact sheets, and our extensive web site- www.als.ca. In addition to being a source of on-line educational materials, the **ALS Society of**

Canada web site is a great place for you to find current, trustworthy information about key ALS research, ALS and Society news, advocacy initiatives, links to provincial Societies and other useful resource sites, and much more. "Knowledge is power", so learn as much as you can!

Providing Support

The ALS Society of Canada and the 10 provincial ALS Societies are here to provide you with information, referrals, and support. Available **services vary by province**, so specific information on what is offered in your province has been provided and included as an additional insert at the end of this Section.

The ALS Society of Canada plays a major role in the production of many **educational publications** that are available to you, including the *Manual for People Living with ALS*. For your reference, a list of ALS Society publications, as well as other educational resources, is included at the end of this manual in the "Educational Resources" section. Contact your provincial Society office to order specific items or go to www.als.ca, where many of them can be downloaded and printed.

Provincial ALS Societies through their affiliated regional offices and/or chapters offer **information** about local resources, **referrals** to community resources that may be of assistance to your individual and family needs, **educational programs** and services such as lending libraries and lectures, **social support programs** such as support groups, and help with acquiring **medical equipment and assistive devices**.

ALS Canada and Provincial Societies are also involved in **special events, public awareness, and advocacy activities** in which you and your friends and family

members may wish to participate such as *ALS Awareness Month* in June, fundraising walks, or advocacy efforts that focus on making changes to public policy to benefit persons affected by ALS. Advocacy activities may involve letter writing campaigns to strategic legislative visits, to a coordinated ALS Day at

federal or provincial legislatures. Participation in Society activities can be very empowering. Contact your provincial ALS Society for information on any upcoming events in your area.

Supporting Research

The ALS Society of Canada is **dedicated to finding the cause of and cure for ALS** through funding quality research. The Society uses a peer review granting process each year to award worthy projects being conducted by scientists in Canada. Since 2000 we have funded research in partnership with Muscular Dystrophy Canada. This Neuromuscular Research Partnership (NRP) works with the Canadian Institutes of Health Research (CIHR) to fund excellent and relevant research.

“ To support research towards a cure for ALS and support ALS Partners (provincial ALS Societies) in their provision of quality support for persons affected by ALS. ”

Only the most promising projects by the finest scientists are funded and, over the years, this strategy has been rewarded with significant advances in our knowledge of ALS. With the acceleration of results due to advances in neurology and other areas of science, we know that, effective therapies and a cure are now, more than ever, within reach.

The ALS Society is planning to launch new opportunities for funding research, including offering studentships, post-doctoral fellowships and young investigator fellowships. Attracting new researchers to the ALS field is an important strategy in meeting the mission to find the cause and cure of ALS.

Read the brief *Research Updates* inserted in the pocket page at the end of the **Resource Section** of the *Manual* to learn about the kind of research the Society is funding. As more updates become available, collect them and keep them together for easy reference.

Contacting the Society

To contact the ALS Society of Canada, please use the information provided below:

ALS Society of Canada
265 Yorkland Blvd. Suite #300
Toronto, Ontario
M2J 1S5
Tel: (416) 497-2267
Fax: (416) 497-1256
1-800-267-4257
website: www.als.ca

Provincial ALS Societies

ALS Society of Alberta (and NWT)

www.alsab.ca
(403) 228-3857
Toll Free: (888) 309-1111

ALS Society of British Columbia (and Yukon)

www.alsbc.ca
(604) 685-0737
Toll Free: (800) 708-3228

ALS Society of Manitoba (and Nunavut)

www.alsmb.ca
(204) 831-1510 or (204) 254-5337
Toll Free (866) 718-1642

ALS Society of New Brunswick

(506) 855-1239
Toll Free: (866) 722-7700

ALS Society of Newfoundland and Labrador

www.envision.ca/webs/alsnl
(709) 634-2435
Toll Free: (888) 364-9499

ALS Society of Nova Scotia

(902) 454-3636
Toll Free: (866) 625-7257

ALS Society of Ontario

www.alsont.ca
(416) 497-2267
Toll Free: (800) 267-4257

ALS Society of Prince Edward Island

(902) 892-7102

ALS Society of Québec

www.sla-quebec.ca
(514) 725-2653
Toll Free: (877) 725-7725

ALS Society of Saskatchewan

(306) 949-4100

YOUR PROVINCIAL ALS SOCIETY

Information about ALS support programs and services available through the ALS Society in your province or territory has been included at the end of this section. Please contact your Provincial Society for further information.

Managing ALS is a continually changing challenge. Although ALS is a degenerative disease, the rate at which neurons and muscles degenerate is unpredictable and varies greatly from one individual to another. In some cases the disease seems to have reached a plateau, while in others it reaches a standstill for varying lengths of time. Also, ALS can progress steadily at a rapid or slow rate. Whatever the rate of muscle degeneration, you should remain as active as possible, without causing fatigue in affected muscles. **It also helps to focus on what you can do, rather than on what you can't do.**

In order to remain as functional as possible for as long as possible, you will need a supportive, patient-centred healthcare team who understands ALS, your personal situation, and the options that will be available to you. As time goes by you will need to work with your healthcare providers to treat various symptoms of ALS and adapt to an array functional changes. An overview of treatments is discussed in this section, but for more information on specific ALS management issues the following sub-section tabs can be found in this section:

- *Adapting to Changes in Mobility and Maintaining Independence*
- *Adapting to Swallowing Problems and Maintaining Good Nutrition*
- *Maintaining Oral Health*
- *Adapting to Changes in Speech and Maintaining Communication*
- *Adapting to Changes in Breathing and Maintaining Lung Function*
- *Approaching End-of-Life Issues and Advance Care Planning*

YOUR HEALTHCARE TEAM

Access to healthcare professionals who are knowledgeable about ALS and work together with you as a team is a very important factor in effectively managing ALS. There are a number of ALS clinics across Canada that care for patients with ALS through an interdisciplinary team approach. You very well may have received your definitive diagnosis of ALS in one of these clinics. In other communities, the same healthcare professionals may be available in private practice or through homecare community programs. Contact information for **clinics and centres in Canada specializing in ALS management can be found on our web site, www.als.ca, or by calling the ALS Society of Canada at 1-800-267-4257.**

Even if you visit an ALS clinic for periodic team follow-up and assessment, you will still need services from healthcare professionals in your community. The ALS specialty team can function as an excellent resource for educating your community practitioners about ALS and your needs. It is important that all of your primary family caregivers meet with members of your healthcare team to be trained in the various helping techniques (e.g., lifting, transferring).

Effective communication between healthcare professionals and their patients is a key factor in getting the best care. To help you get the most out of your healthcare visits, see tips provided by Health Canada at http://www.hc-c.gc.ca/english/feature/magazine/2000_10/doc_e.htm. Click on the communication skills tools link and view or download and the brochure, It Helps to Talk.

Primary Doctor

Your doctor's tasks may include the following:

- Explaining the diagnosis and possible progression of ALS;
- Making necessary referrals to, and consulting with, other healthcare providers to best manage your care;
- Discussing course of action you wish to take if respiratory failure occurs

Neurologist

Your doctor will refer you to a neurologist, a specialist in diseases of the nervous system who can confirm a diagnosis of ALS. Between them they will:

- outline types of treatment options available;
- encourage the setting of short-term goals;
- help you preserve a positive self-image and maintain your morale;
- work with you to identify specific needs and concerns and refer you to therapists who may be able to find solutions.

Nurse Clinician

A nurse clinician generally formulates nursing goals after analyzing assessment data, then draws up and implements a care plan. Other nursing functions may include the following:

- explaining terminology and techniques;
- teaching skills and providing demonstrations;
- evaluating skill levels and reviewing procedures;
- addressing questions and concerns;
- ensuring the comfort of people living with ALS;
- encouraging and promoting decision-making by the people with ALS and their families;
- liaising with community organizations;
- making referrals;
- limiting the development of complications;
- providing nursing interventions if problems arise.

Occupational Therapist, Physiotherapist, and Physiatrist

The occupational therapist (OT) and the physical therapist/physiotherapist (PT) develop strategies that enable people with ALS to continue to carry out daily activities in a safe and efficient manner. This is accomplished through the following:

- teaching by demonstration;
- ongoing assessment/evaluation;
- ordering appropriate equipment in a timely manner;
- preventing complications;
- utilizing community resources;
- providing psychological support.

The OT is generally more concerned with fine-motor control and how movement will affect function. The OT's tasks may include the following:

- assessing fine-motor functional abilities (for example, hand use);
- evaluating positioning and seating requirements;
- assisting in choosing and accessing suitable augmentative mobility and communications strategies and any required assistive devices;
- giving instruction to the person with ALS, family members, and caregivers on exercise, positioning, correct body mechanics for lifting and transferring, and use of assistive devices;
- teaching energy conservation and time management techniques;
- providing information about modifications to home and other environments to enhance mobility and safety.

The PT is generally more concerned with gross-motor function and anatomical impairments associated with abnormal movement. The PT's tasks may include the following:

- providing a detailed analysis of abnormal movement (for example, gait analysis-how you walk);
- optimizing the person's strength, function, and comfort;
- designing and monitoring a therapeutic exercise regimen when appropriate;
- assisting with breathing management;
- providing training in energy conservation and time management techniques;
- preventing needless purchases of unsuitable equipment;
- making a home assessment.

Access to healthcare professionals who are knowledgeable about ALS and work together with you as a team is a very important factor in effectively managing ALS.

The physiatrist, a medical doctor specializing in physical and rehabilitative medicine, generally evaluates the extent of disability and functioning, and gauges the level of muscle function you still have (residual level). On the basis of these findings, treatment plans may be designed. Other functions may include:

- recommending preventive and supportive treatment;
- preventing complications;
- determining which diagnostic tests are necessary;
- helping maintain maximum function and quality of life;
- consulting with therapists concerning orthoses and equipment.

Dietitian

A dietitian's primary considerations are to keep your quality of life as high as possible by maintaining safe and adequate nutrition and hydration, in order to prevent life-threatening nutritional deficits from developing. The dietitian may assess or evaluate the following:

- functional abilities (ability to self-feed or to administer tube feedings);
- nutritional status and present intake.

The dietitian may recommend the following strategies:

- appropriate changes in food texture and consistency;
- appropriate methods of food preparation;
- substitutions for hard-to-manage foods;
- meals of a manageable size and frequency;
- strategies for maximizing nutritional intake.

Respiratory Therapist and Spirologist

The respiratory therapist generally devises strategies to optimize remaining muscle function and reduce discomfort, and institutes a program of chest care if necessary. In some locations, the physiotherapist may be the person who helps you with exercises to promote airway clearance and cough techniques. Other tasks may include:

- evaluating pulmonary function status;
- maintaining pulmonary hygiene (airway clearance);
- providing suggestions for managing decreasing breathing function;
- offering information on body positioning, energy con-

servation, relaxation, and compensatory techniques to improve breath support for nutrition and for speech;

- setting up a home ventilation program if appropriate;
- making suggestions about a course of action to take when respiratory failure occurs.

The respirologist is a medical doctor who specializes in care for the respiratory system. During the course of ALS you may be referred to or seen by a respirologist in the event of a respiratory infection, or when you decide you are interested in mechanical ventilation.

Speech-Language Pathologist (SLP)

A speech-language pathologist provides advice on techniques and strategies to allow the person with ALS to continue to communicate throughout life. Some functions of the speech-language pathologist may include:

- evaluating the individual's motivation and potential for learning new techniques;
- evaluating functional abilities, such as oral motor function, cognitive-linguistic function, augmentative communication function, and swallowing function;
- determining the most efficient communication function;
- training people with ALS and family members in techniques of effective communication and energy conservation, and safe eating, drinking and swallowing techniques.

Social Worker

In addition to other healthcare professionals, you may be referred to a social worker. The social worker's tasks include both practical assistance as well as emotional support to help people with ALS and their families cope with everyday life. The social worker offers:

- emotional support and counselling to the person with ALS and to family members and refers to the appropriate service agency if ongoing support is required;
- an understanding of the emotional impact of an ALS diagnosis and assists with the psychological adjustments that come with this diagnosis;
- information about available community resources and acts as an advocate to assist people in accessing these resources;
- information on legal and financial issues and assists people to access these resources in their own communities; and

- help with setting short-term and long range goals and making plans that will meet future care needs.

Pastoral Care Worker

A minister, priest, rabbi, chaplain or other pastoral care worker may perform the following functions:

- listening to and empathizing with those who want to vent concerns;
- assisting in making decisions;
- giving spiritual support during emotional or physical crises;
- acting as an advocate for those who have no voice;
- reassuring people with ALS that their lives have meaning and facilitating spiritual reflection;
- encouraging people with ALS to discover their personal strengths;
- encouraging people with ALS to recapture positive experiences from the past by recall or reminiscence;
- celebrating the individual's humanity and worth;
- listening to concerns about death and dying; and
- arranging for and conducting services when appropriate.

There may be other healthcare professionals on your ALS team such as an assistive technology expert, wheel chair seating expert, psychologist, or gastroenterologist. The above mentioned professionals comprise the core members of an interdisciplinary ALS care team.

A business card holder page has been included in the *Manual* to help you keep the contact information for your healthcare team members handy when you have questions or need to make appointments.

Professional Associations

To learn more about some of the professions described, contact the professional associations listed below:

Canadian Association of Occupational Therapy

Phone: (613) 523-CAOT (2268)/ (800) 434-CAOT (2268)

Fax: (613) 523-2552

www.caot.ca

Canadian Association for Pastoral Practice and Education

Phone: (902) 820-3085 (Halifax)/(866) 442-2773

Fax: (902) 820-3087

E-mail: office@cappe.org

Canadian Association of Social Workers

Phone: (613) 729-6668

Fax: (613) 729-9608

E-mail: casw@casw-acts.ca

Canadian Association of Speech-Language Pathologists and Audiologists

Phone: (800) 259-8519

Fax: (613) 567-2859

E-mail: caslpa@caslpa.ca

Canadian Physiotherapy Association

Phone: (416) 932-1888 or 1-800-387-8679

Fax: (416) 932-9708

E-mail: information@physiotherapy.ca

Canadian Society of Respiratory Therapists

Phone: (613) 731-3164/ (800) 267-3422

Fax: (613) 521-4314

E-mail: csrt@csrt.com

Dietitians of Canada

Phone: (416) 596-0857

Fax: (416) 596-0603

www.dietitians.ca

OVERVIEW OF ALS DISEASE MANAGEMENT ISSUES

Although there are no known medications that will cure ALS, there are medical treatments, therapies, and assistive devices which help people maintain active and independent lives for as long as possible. These **disease management and adaptive strategies promote the well-being of both you and your family.** Communicate openly with your caregivers, therapists, and doctors about your symptoms and challenges so that you can work together to find solutions that work best for you in a timely manner.

Some of the problems associated with ALS and the strategies for managing them are discussed in more detail in the additional topic specific sub-sections. When you decide you are ready to learn more about these topics or you are experiencing problems in these areas, the information will be available for you.

Mouth and Throat Problems

- **Speaking and Swallowing**

As mentioned before, when the onset symptoms of ALS are speaking or swallowing problems, it is known as Bulbar ALS. For other people with ALS, these problems don't generally occur until the later phases of the disease.

People with Bulbar ALS are vulnerable to significant weight loss, and should consider a feeding tube in the earlier phases of the disease. Generally speaking, when you have lost 10-15% of your "normal" body weight, your doctor should talk to you about the option of a feeding tube while you are fairly healthy.

For those who do lose their ability to speak, there is now a range of communication aids available from low-tech to very high-tech.

See [Adapting to Swallowing Problems and Maintaining Good Nutrition](#) and [Adapting to Changes in Speech and Maintaining Communication](#) sub-sections.

- **Choking**

Those who develop swallowing problems are at risk for choking. Because this could become dangerous, get advice from your doctor and other healthcare professionals about how to address this problem. There are things you can do regarding how you eat and what you eat that can lower the risk of getting food stuck in your throat. If you are beginning to notice problems with swallowing, see your doctor for a referral for a swallow study.

Choking is an alarming symptom. Many people assume that choking to death is how people with ALS die. This has been shown to be false. Choking is a manageable symptom, and most people with ALS die peacefully.

See [Adapting to Swallowing Problems and Maintaining Good Nutrition](#) sub-section.

- **Build-up of Saliva and Drooling**

People with Bulbar ALS often develop an excessive drooling problem because they have difficulty swallowing the normal production of saliva. An increase in the actual production of saliva may occur as a side-effect of tube feeding diets. Your doctor can offer various medications to limit saliva production to reduce this problem.

See [Maintaining Oral Health](#) sub-section.

- **Saliva and Mucus in the Lungs**

Should muscles involved in breathing (diaphragm) and coughing (abdominal and intercostals) weaken, it becomes difficult to cough up saliva and mucus. If you are having this problem, see your doctor as soon as possible. There is a technique for freeing saliva and mucus from the throat and lungs, which involves lying on a slope with your head on the down side so that the saliva flows down when the technique is applied. This technique should be taught by a trained professional who is familiar with and can demonstrate the correct position and procedures. This is especially important if you are experiencing shortness of breath. Also, a suction unit can help remove saliva from the mouth and prevent it from flowing back into the lungs.

See *Adapting to Changes in Breathing and Maintaining Lung Health* sub-section.

- **Coughing**

Coughing can occur for more than one reason. For example the lungs can clear a passage by automatically and unexpectedly forcing out air, while at the same time irritations in the throat due to blockage can also cause unexpected coughing. People with ALS may develop weak coughs due to breathing muscle weakness which makes it difficult to effectively clear airways that may be blocked. Talk to your doctor or respiratory therapist about techniques to produce an effective cough. If you do not have a respiratory therapist, your physiotherapist may be able to assist you.

See *Adapting to Changes in Breathing and Maintaining Lung Function* and *Adapting to Swallowing Problems and Maintaining Good Nutrition* sub-sections.

Symptoms that Affect Daily Living

- **Eating**

Continue to feed yourself as long as possible. However, if you feel too weak to manage a whole meal by yourself, or have trouble with muscle control to operate knives, forks and spoons, ask for help from your caregiver. For instance, your caregiver could help a lot just by cutting your food for you. An occupational therapist can also help by showing you how to adapt your eating utensils so they are easier to grip, or adjust the height of your eating surface so you have less distance between the plate and your mouth.

When swallowing difficulties arise, or jaw muscles involved in chewing are weakened or stiff, your dietitian will be able to help you with how to eat, what to eat, and how to modify food textures to make eating easier and safer. When difficulty with eating is accompanied by weight loss, you should talk to your doctor about the possibility of tube feeding.

See *Adapting to Swallowing Problems and Maintaining Good Nutrition* sub-section.

- **Constipation**

Constipation may occur as a result of inactivity or lack of adequate fibre or water in the diet. For those using tube feeding, it may be especially difficult getting sufficient fibre and hydration. In either case, if you are experiencing constipation, ask a dietitian about ways to add more fibre to your diet. If adding fibre to your diet doesn't solve the problem, see a doctor about an appropriate medication.

- **Fatigue**

Fatigue occurs as a result of weakening muscles, suspected higher metabolic functioning in ALS patients, and declining breathing function. One of the first ways you can combat fatigue is to conserve your energy for really important tasks or activities that you really enjoy. An occupational therapist can plan a daily routine with you that will help you to adapt to life with ALS. Some tasks can be done in different ways that will save some of your energy. There are a large num-

ber of aids that can help you to do the things you now find difficult. It is essential that you consult your therapist before buying an aid to avoid expensive mistakes.

If you tend to wake up in the morning feeling very fatigued and light headed, discuss this with your doctor promptly as you may not be breathing well enough in the night, thus requiring a respiratory function evaluation and some airway management strategies.

- **Getting to Sleep and Positional Problems**

Some people with ALS may become immobile. People who are unable to move get very uncomfortable lying in the same position while sleeping for several hours. There are special beds, which help an immobile person sleep without being manually turned during the night. A satin bottom sheet and nightwear also facilitates turning.

Getting to sleep can also be problematic for some people. Talk to your doctor about relaxation exercises you could use to try to get to sleep. If medication is required to help you sleep, be mindful that use of sedatives and tranquillizing medication depresses respiration and should be used cautiously by those who may have impaired pulmonary function.

See Section 5 *Assistive Equipment* section and *Adapting to Breathing Changes and Maintaining Lung Function* sub-section.

- **Use of Alcohol and Medication**

If you are taking any medications, be very careful about drinking alcohol. The combination of alcohol and many medications can cause serious problems. This is not always predictable. For example, alcohol with some medications, such as a simple cough remedy, even taken several hours apart, can restrict a person's ability to breathe, and at the same time cause a coughing spell, both lasting for hours. Also, reduced motor control can be accentuated by alcohol. If you like to have the occasional alcoholic beverage, talk to your doctor about the specific risks that may be involved.

- **Excessive Emotional Outbursts**

Uncontrollable emotions such as excessive laughing or crying may be experienced by some people with ALS. This is called emotional lability, or pseudobulbar affect, and can be particularly frustrating because the outburst is often caused by something very trivial and may be misunderstood by other people. This symptom usually catches people by surprise when it first happens; however, over time, many learn how to modify these emotional outbreaks and how to avoid some of the situations in which they occur. If this is happening to you, talk to your doctor about medications that may help control this problem. People with pseudobulbar affect may be helped by antidepressant drugs.

- **Cognitive and Behavioural Changes**

Until fairly recently, it was assumed that ALS did not affect thinking. However, researchers have found that changes in the way one thinks, perceives, and processes information (cognition), and behaviours will occur in some patients with ALS and sometimes early in the disease before an ALS diagnosis has been made. This is not to say these changes will occur in all persons with ALS and the exact nature of the changes can vary from person to person. You and your family can better identify any such changes and discuss effective management strategies with your ALS healthcare team members if you are educated about them. Proper management may lessen the impact of cognitive and behavioural changes on daily living and coping with ALS.

The following may indicate changes in thinking:

- Reduced reasoning, insight, and problem-solving ability
- Deficits in verbal fluency (reduced word recognition and word choice)

Some of the following symptoms of personality and behavioural changes may be experienced:

- Apathy
- Loss of inhibition
- Restlessness or overactivity
- Social inappropriateness
- Mood swings
- Compulsive rituals such as repeatedly dressing, using the bathroom, eating, hoarding, etc.

For more information about cognitive, personality, or behavioural changes in ALS, consult with an ALS specialist.

Mobility

- **Joint and Muscle Pain**

Persons with ALS who lose significant muscle function in areas that involve joints may end up not using those joints to their full capacity. Lack of use can cause stiffness and joint pain. **Range-of-motion exercises are designed to prevent these joint problems.** Careful attention to your exercise regime, whether active (doing it yourself) or passive (with assistance), will eliminate much potential joint pain. However, there are still a number of common pains that can develop. If your arms are weak and you allow them to hang unsupported from the shoulder, there is a tendency for the shoulder joint to become painful. It is helpful to support weak arms whenever possible on pillows, armrests or on a table.

A shoulder sling will also give the arm some support and decrease strain on the shoulder joint while you are walking. Hip pain can result from prolonged sitting in a sagging seat or chair. A firm seat on a regular chair or wheelchair will relieve strain on the hip joints.

- **Leg and Foot Swelling**

If your lower limb mobility is reduced, you may experience mild leg and foot swelling, which is best reduced by moving the toes and ankle, if possible, and by elevating the leg and/or using an elastic stocking. Talk to a nurse about how best to address this problem.

- **Muscle Cramping**

Cramps are not uncommon in people with ALS. They can be alleviated to some extent by keeping the affected muscle warm and by stretching it or having your caregiver stretch it until the pain is eased. Severe or frequent cramps should be discussed with your doctor. There are a number of medications available to reduce cramping.

- **Changes to Posture**

If the muscles that maintain your upward posture weaken, you may have discomfort in your lower back, neck and shoulder blade region. Special cushions, chair backs, lumbar (lower back) and cervical (neck) rolls are available to help you to maintain correct sitting posture. It may be necessary for you to sit in a reclined position or to use a neck collar to maintain proper positioning. Your occupational

therapist can assist you in choosing the right device.

- **Difficulty Walking**

If you experience weakening leg and ankle muscles and unexpected fatigue, tripping and falling become problems. When these problems occur you should consider using leg and ankle splint devices (**ankle foot orthoses-AFOs**) that provide added support to weakened muscles, or a cane or walker as soon as you are threatened by unexpected falls. Some people are self-conscious about having to use walking aids, or see it as an announcement that their condition is worsening. These are completely normal reactions. However, you must also think about preventing injuries that will inhibit your independence further, or cause serious harm to you such as a head injury.

- **Difficulty Gripping and Holding**

Some people with ALS lose strength in the hand and wrist muscles, losing the ability to manage small hand movements such as holding a pen to write, or cutlery to eat, turning a key to start a car, or turning a handle to open a door. There are a variety of products designed to assist you with weakened grip strength.

If these weaknesses continue to develop, you can lose your ability to hold even lighter weight articles. Getting dressed and undressed becomes more and more difficult, and then impossible to do alone. As this process occurs, dressing can be made easier with Velcro fasteners, elastic waistbands, and other features that make clothing easier to put on and take off.

Cold or hot hands can weaken the grip of most people. For persons with ALS, however, this is even more pronounced. If your fingers get cold you will probably be surprised by how much harder it is to do such things as undo buttons, or turn a doorknob. An occupational therapist will be able to help you with specific strategies. Some OTs specialize in hand movement and therapy.

As ALS progresses, there are various types of equipment available to assist you with your mobility needs. With respect to moving about, some may need a cane or a walker at first, then a manual wheelchair, followed by a power wheelchair to assist in maintaining a certain level of independence. Also, there are raised chairs and toilet seats which are easier to get up from, and portable hoists to move a person around.

ALS. Three letters that change people's lives. FOREVER.

Regarding eating, there are eating utensils that can assist and increase independence - cutlery with large easy-grip handles, non-slip mats and special plastic plates.

Choosing the best assistive equipment for your situation involves many considerations, including an understanding of longer-term needs. It is advised that you get advice from a healthcare professional such as an occupational or physiotherapist before acquiring assistive equipment.

See Section 5 *Assistive Equipment* and Section 4 *Adapting to Changes in Mobility and Maintaining Independence* sub-section.

Sexual Concerns

Sexuality and intimacy are basic aspects of human life. Sexual desires and abilities may not be affected by the disease process, except for the physical limitations imposed by physical discomfort, muscle weakness, fatigue or low energy levels, or medication side-effects. A person with ALS may worry about not being able to please a healthy partner. For men, anxiety may cause impotence.

Other factors that may contribute to unsatisfactory sexual relations are:

- Adjustment to assistive devices or support systems
- Dealing with everyday survival
- Negative self-image
- Reduced independence with self-grooming
- Reduced communication ability
- Limited mobility
- Changes in physical appearance
- Altered role
- Emotional state
- Functional level

You and your partner may want counselling with an empathetic professional to openly deal with mutual concerns and expectations. If you are both willing, you could explore different sexual techniques, role flexibility, and alternative methods of sexual expression.

The following suggestions may prove helpful in dealing with sexual concerns:

- Using techniques, assistive devices, and positioning to accommodate increasing muscle weakness and other

symptoms of ALS

- Identifying techniques and assistive devices that enable you to maintain good grooming and personal cleanliness
- Maintaining communication (the open expression of affection and need is important.)
- Respecting the boundaries of the other partner
- Altering the living environment to provide adequate privacy and reduce embarrassment
- Scheduling of "adult time" if there are young children in the family
- Wearing street clothes rather than night attire whenever possible during the day to emphasize normality and reduce playing the sick role
- Re-focusing interests and energies into other areas

Partners should realize that touching is as important as sexual performance in reducing tension and maintaining emotional intimacy. Preservation of personal integrity should be an overriding concern. If you have any questions or concerns, speak with your doctor and request a referral to a sexual health clinician in your area.

Complementary and Alternative Healthcare (CAHC) and Natural Health Products (NHP)

In the search for better health and well-being, the healthy as well as those managing an illness for which there is no cure or limited treatment often look to complementary and alternative health care (CAHC) and natural health products (NHPs) for answers.

Complementary and alternative healthcare (CAH)

refers to practices that complement mainstream medicine by contributing to a common whole, by satisfying a demand not met by conventional approaches.

Massage and meditation are CAH practices that have become quite common in chronic disease management.

Natural health products (NHPs) refer to herbs, vitamins, minerals, essential fatty acids, and

homeopathies.

It is also recommended that you and your doctor read **Fact Sheet #10--ALS/MND: The Dangers of Unproven Therapies,** written by Dr. Andrew Eisen and **Fact Sheet #11-Natural Health Products-Making Sense of Complementary and Alternative Medicines**

both included in the last section of the Manual.

There is very little scientific evidence to support the safety and effectiveness of many CAHC and NHP claims and in some cases may be detrimental to a person with a disease such as ALS, or anyone on any medication.

However, there are certainly legitimate practices, such as massage and supplement use (e.g., vitamin E) that may have a positive effect on feelings of well-being. The ALS Society is supportive of you and your doctor working together to find what works best for you. **If you are seeing an alternative practitioner, let your medical doctor know as you need to take care that nothing interferes with the medicines you are taking to manage ALS symptoms.**

No matter how desperate you are to find a cure for ALS, be very wary of unconventional treatments that make claims of being able to "cure" or "reverse ALS." Usually the people offering these "cures" charge thousands and thousands of dollars leaving families left with no money to pay for essentials or for needed traditional symptom management treatments. Unfortunately, many of the people pushing these claims are making a lucrative living at the expense of people who are looking for hope.

The following information provided takes a look at massage, meditation, and NHPs in general terms.

- **Massage and Touch Therapies**

Touch therapies can be very relaxing and comforting for people with ALS. They can warm up limbs, which often feel cold because of inactivity, low circulation and decreased muscle mass. Though touch therapies are usually given by a caregiver or therapist, self-massage can also be very beneficial. It is important to talk to your doctor before receiving massage therapy.

Caution: While having ALS is not a contraindication to massage therapy, modifications in technique may be required and should be done so under medical consultation.

The easiest touch therapy to learn is massage. For the most beneficial massage, body oil is used to lubricate the skin, which aids in soothing and relaxing the muscles. Massage involves systematically stroking, kneading and pressing the soft tissues of the body with fingers and the palm of the hand, working on the muscles, ligaments and tendons. A complete massage covers the entire body, inducing a state of warmth and relaxation. However a massage of parts of the body, such as arms, legs or shoulders is also beneficial and will help improve circulation.

Shiatsu:

Shiatsu is a Japanese system of touch therapy and is given using fingers, thumbs and hands. The forearm, knees, and feet can also be used, applying pressure to specific points on the body.

Reflexology:

Reflexology is based on the theory that pressing and massaging certain points in the feet will affect the whole body and induce relaxation.

Aromatherapy:

Aromatherapy is a massage that involves the use of fragrant oils, which penetrate the skin, adding another sense of pleasure to relaxation.

Cranio-Sacral:

A cranio-sacral massage is a very light and gentle massage of the back of the head and neck. It is non-invasive and very soothing.

Tens Machine:

A Tens Machine stimulates muscles through electronic impulses, and is often used by physiotherapists. The cost of most massage treatments ranges between \$40 and \$75 for sessions up to an hour in length. Most of these treatments are not covered by provincial healthcare plans, but may be covered in whole or in part by private or group insurance if they are prescribed and/or delivered by a registered therapist.

- **Meditation**

Meditation is a way to relax and revitalize both mind and body, and can be practised in many ways. Technically, meditation is deliberately entering into an inner silence and stillness for a certain period of time, usually 15-20 minutes. It has been practised over the centuries by many different cultures and in many different ways. Referring to the results of researching meditation in his book *The Relaxation Response*, (Avon Books, N.Y.), Dr. Herbert Benson writes that meditation creates an overall awareness of relaxation and a feeling of well-being. Breathing and heart rate slow down, blood pressure drops, and there is an increase of healing alpha waves from the brain. In other words, meditation can reduce stress, and relax muscles.

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Meditation techniques can be divided into two categories. One type of meditation involves concentrating on something, such as your breathing, a candle flame, a religious object, or even rolling surf, while peacefully and silently repeating a mantra, a special word or sound. Any word that contributes to inner peacefulness can work. The other type of meditation also involves repeating a mantra, but the objective is to relax your mind entirely. When thoughts come into your mind, you try to relax, observe the thoughts and let them flow, without participating in the thought process. For most people, this takes practice.

Before starting to meditate it is usually best to let other people in your house know that you don't want to be disturbed for 20 minutes. Many meditation teachers suggest that, for the best results, meditation should be done twice a day, for about twenty minutes, with your eyes closed. When you think about 20 minutes is up, gently open your eyes and look at your watch. Two common problems for those new to meditation are being unable to relax as their minds are so full of thoughts and/or when they are finally relaxed, they fall asleep.

It takes daily practice and time to develop the ability to achieve a state of conscious relaxation. Sometimes quiet, peaceful music can help the mind calm down, but in later phases of meditation that may be too distracting.

There are several other techniques to help clear the mind, and achieve a state of relaxation, including music therapy and visualization therapy. Books on this subject can provide patients with knowledge of these techniques, instructions, exercises and many hours of quiet pleasure. Your nurse, social worker, occupational therapist, or other mental health professional may also be good resources for information and training.

- **Natural Health Products (NHPs)**

Because vitamins, herbs and other so-called complementary or alternative medicines (natural health products) have become so popular over the last few years, it is appropriate to say a few things about these potential remedies in the context of research. People with ALS can be particularly attracted to these remedies either because drug trials so far have been unsuccessful, or because of the perception that alternative medicines are natural.

ALS. Three letters that change people's lives. FOREVER.

While extensive and rigorously controlled research is done for pharmaceutical drugs before they can go to market, much, much less research is done on NHPs with the exception of vitamins, which have been extensively researched. On the other hand many NHPs have been used by some cultures for hundreds and even thousands of years. Stories circulate about people who have been cured by these medicines. Also, it is well known that animal and plant by-products have provided the basis for many successful drugs. Unfortunately, the hundreds of millions of dollars required to thoroughly test pharmaceutical medicines are not available to test alternative medicines. Scientifically, little is known about the effect of alternative medicines on the human body.

New Regulations in Canada:

As of January 2004, new NHP Regulations under Health Canada, designed to balance the need for safety and efficacy with consumer choice. The new regulations stipulate that every NHP sold in Canada must be issued a product licence, which registers it with Health Canada. The new regulatory framework incorporates an evidence-based approach that assesses products for safety and effectiveness.

For more information on natural health products, see **Fact Sheet #11--Natural Health Products.**

The **Standards of Evidence** developed are clearly defined criteria concerning the amount of

evidence required to support each claim (five levels ranging from "well-designed systematic reviews and meta-analysis of randomized controlled trials (RCT) or at least one well-designed RCT" to "references to traditional use"). The stronger the claim, the stronger the supporting evidence needs to be. As a consumer, you will now be able to tell what level of evidence is available which will help inform your choice. Talk to your doctor about the claims a product is making in terms of what it says it can do for you. Your doctor can help you decide if it could be harmful or could negate the effect of other medications you are taking. Also consider the cost of the product. If it is very expensive, but there is little if any scientific evidence to support the claims, you may want to reconsider.

If you find something that helps you feel better or makes your life a little bit easier, let your doctor know about your progress. He or she may want to study it further, or share it with other appropriate patients.

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Everyday activities, such as getting out of bed, sitting down to dinner, carrying in bags, are often taken for granted. As ALS progresses, and motor neurons die, these simple actions can become more difficult and require more effort. Working with an occupational therapist and a physiotherapist familiar with ALS will help you come up with ways to stay as functionally mobile as you can to continue to accomplish the everyday activities you are accustomed to doing.

Mobility strategies include exercise and mobility aids. These strategies aim to promote physical comfort, prevent injury, and help maintain independent living. Caregivers will also benefit from mobility strategies in terms of injury prevention.

EXERCISE AND ALS

As motor neurons die, muscles become weaker and stiffer. As a result, someone with ALS will become less mobile in terms of moving their head and limbs. When it is difficult to move body parts, the associated joints become stiff which can be quite painful.

Therapeutic exercise can help relieve discomfort. Recreational exercise can also help with physical symptoms as well as boost one's enjoyment of living.

It is important to realize that exercise will not strengthen muscles that have been weakened by ALS. Once the supply of motor neurons that control a particular muscle has degenerated, it cannot be regenerated by exercise or anything else.

The purpose of exercise for people with ALS is:

- To maintain or improve the flexibility of muscles not affected by ALS
- To maintain the flexibility of muscles that have been affected
- To maintain the flexibility of joints in the neck, trunk and limbs

Range-of-Motion (ROM) Exercises

A person with ALS needs to move each affected joint through a series of range-of-motion (ROM) exercises

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every day to prevent joint stiffening. Exercise will help to keep your body as flexible as possible and your joints mobile. ROM exercises are usually done systematically, meaning that the joints of one limb are exercised in a particular order before the next limb is exercised and so on.

The objective of ROM exercises is to move each affected joint through its full range of motion every day. Not every person with ALS can do a full set of active exercises.

An **active exercise** is one you do yourself without any assistance, when your muscles can perform the full movement. When you cannot move through a ROM exercise on your own, you can still complete the movement as an **active-assisted exercise**. A helper may assist the muscle through the movement, or you may be shown a way to do a self-assisted range of motion exercise. **Passive exercises** are done completely by a helper when muscles can no longer perform

Mobility strategies... aim to promote physical comfort, prevent injury, and help maintain independent living.

any of the movement. The helper moves the joints through their ROM by manipulating your limb. Passive exercises work the joints but not the muscles. Your therapist can train your caregiver(s) to do these exercises properly.

The transition from active to passive exercise is seldom abrupt. You may find that you can do some exercises actively, some with assistance and still others only passively.

Each person with ALS needs an exercise program tailored to their own individual needs and abilities. Your doctor and physiotherapist can prescribe the exercises that are right for you at any given time. Your physiotherapist will demonstrate the exercises and ensure that they will be performed correctly. Ask for diagrams of the exercises to help you and your caregiver remember the techniques.

It is important that all exercise be performed in moderation. Fatigue will only increase your weakness and rob you of energy that you need for your daily routines and the activities you enjoy. If you find that your prescribed set of exercises tires you, talk to your therapist. Changes can be made that will eliminate the risk of fatigue.

Similarly, none of your exercises should cause pain. If you do experience pain when exercising, stop that exercise and talk to your therapist. It may be that you are not doing the exercise correctly, or perhaps some modification to your exercise program must be made.

Recreational Exercise

If you enjoy such activities as walking, stationary bicycling, and especially swimming, **keep them up for as long as you can do them safely.** If you experience cramping or fatigue, do not continue the exercise until you have consulted your doctor or therapist. Your physiotherapist and occupational therapists will help you make adaptations to both the activity and the equipment (if involved) to help you continue these activities even as your abilities change.

MOBILITY AIDS

Mobility aids are used to:

- Prevent injuries
- Promote independent mobility

Injury Prevention

As limbs become weakened, stiff, or easily fatigued mobility aids should be discussed with your doctor. Appropriate use of mobility assistive devices will **reduce your risk for falls.** However, sometimes falls are the trigger for someone to ask their doctor or physiotherapist about mobility aids.

In the event that you find yourself falling, it is better to drop straight down, and not fall forward or backward to avoid a head injury. The best way to get up from a fall depends on what muscles you can still use. If you still have enough arm and hand strength, you can hold on to something firm, such as furniture, for example and pull yourself up and into a chair.

Some Exercising Tips...

Consult with a physiotherapist to design a program. **Exercises should be done daily and should become a routine.** You may wish to break up your exercise routine into parts to avoid fatigue. If you experience fatigue, consult your therapist for a change in your program.

Do as many active exercises as you can. It may be necessary to switch to active-assisted or passive exercises later. Your therapist can help you to make decisions about the correct limits of your exercise.

Prioritize your exercises based on how effective they are for you in terms of injury and pain prevention and maximizing the functions most important to you. Ask your therapist which exercises are the most important ones to do if you have a busy day ahead of you to conserve energy.

Find out which exercise position is best for you. Some exercises can be done while you either sit or lie down. Passive exercises are usually performed while you lie down. Your therapist can advise you on best positions.

Stop doing any exercise that hurts, and consult your therapist if this happens.

When you have fallen, the most important thing to do is to get help to sit in an upright position. The level of assistance needed will depend on the level of muscle weakness. You may only need a little support while rising, or you may need two people to assist you into a chair or wheelchair. It is important that caregivers not strain themselves, but make you comfortable until additional help is available. **Ask your therapist to teach you and your caregivers the best method of recovering from falls.**

Joint pain and stiffness injuries occur when you are unable to move yourself and you spend too long in one position. This can be very uncomfortable for both the skin and the joints. Arrange for your caregivers to change your position every couple of hours throughout the day and to turn you at night. Some people with ALS improve their comfort in bed by

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using a sheepskin, egg crate foam, a satin bottom sheet or a vibrating air mattress. Your nurse or therapist can discuss the options with you and help you to decide what to try.

If you experience joint pain, discuss this with a doctor or physiotherapist.

Injuries to both persons with ALS and caregivers can occur during transfers. All your caregivers should be instructed in safe and effective transferring techniques by professional healthcare providers as soon as possible. It is important that proper body mechanics be taught to decrease the risk of injury to both you and your caregiver. In addition to body mechanics, there are transfer devices such as boards and lifts that can be used to prevent injury.

Assistive Devices

Assistive devices range from walking aids to transfer devices to home adaptations such as ramps. Additional information on assistive devices is included in the manual under the *Assistive Equipment* section.

- **Canes and Walkers**

Most ALS patients will require a cane or walker sooner or later. The choice of a cane or walker should be made in consultation with your doctor and/or physiotherapist. Canes and walkers will provide the stability you need while you are still able to walk on your own without sever fatigue. Walkers provide more stability than canes.

There are several types of canes: single point (straight cane with a handle); four-point (a cane with a four pronged bottom); and Canadian crutches (a crutch not positioned under the arm, but has a section that grips onto the forearm and has a hand grip).

Walkers come in many forms. Some of the cheaper, standard issue types may not be the best choice for someone with ALS. Tripod walkers are also to be avoided. **Types of walkers that may be considered**

by you and your therapist are: two-wheeled, non-swivel (four-legged with two front wheels that do not swivel); two-wheeled swivel (front swivel wheels help with turning); two-wheeled with brakes (front or rear pushdowns to brake if hand weakness present); and four-wheeled with brakes (front swivel wheels and hand or pushdown brakes).

- **Orthoses**

Orthoses are devices that are attached to your body to support weak joints as well as aid in positioning and contracture prevention when there is spasticity. Orthoses help to maintain function and comfort. The most commonly recommended devices for people with ALS are ankle-foot orthoses (AFO), cervical (neck) collar; low-back brace; and resting hand splint.

With respect to moving around on foot, AFOs are customarily recommended if you experience "foot drop." Weakened ankle muscles cause difficulty picking up the foot to walk properly without tripping. Your body alone may compensate by utilizing the thigh muscle more and more to lift the foot high enough to avoid tripping. However, this can cause over-fatigue. Use of braces will reduce the need for your thigh muscles to do all the work, thus enhancing endurance and reducing tripping and consequent falls.

“Treat your equipment as helpful friends, not objects of immobility. I have named my walker "Mickey" and for my wheelchair I am thinking of "Boudicca" for the chariot that carried Queen Boudicca into battle when she almost chased the Romans out of Britain in 62 AD.”

**Bobbi Harris, 84
Person with ALS**

Your doctor usually prescribes such devices, and your therapist will fit and instruct you in proper use of the aid.

- **Wheelchairs**

Many people with ALS will find themselves in need of a wheelchair at some point in time. Some will use it only for long excursions outside the home, some for activities only, and some people will spend most of their day in a wheelchair. When and how often you need to use a chair and the type of chair you need will change overtime.

The decision about when to acquire a wheelchair is one that you will make with your doctor and/or occupational therapist.

Questions you need to answer include:

- Do you always need another person to help you to rise from a seated position?
- Do you stumble and fall a lot?
- Are you fatigued when you walk?
- Do you avoid outings because you are afraid of injuring yourself?

No one is eager to use a wheelchair and this is natural, but a wheelchair should be viewed as an opportunity to increase your independence and your ability to get around. It will also help you to conserve your energy. A therapist can help you decide on and obtain the most appropriate chair.

Selecting a chair will depend on your own personal needs which will require much input from an OT, PT, or rehabilitation specialist to assess your needs and determine required customization for fit and accessories. Because ALS is progressive, you may need different types of wheelchairs for varying periods of time. Try to borrow some of the types of chairs you may not need on an ongoing basis to save your resources for more expensive equipment later on.

Wheelchairs range in type with the major two categories being manual and power. Manual chairs are much less expensive and are most appropriate for someone who still has some ability to move themselves. Power chairs are most appropriate for someone who cannot. Before purchasing a power chair you need to make sure your home is wheelchair accessible and it can be taken apart to fit in a vehicle.

A typical manual wheelchair for someone with ALS may include the following features: high reclining back with headrest; elevating leg rests; seat cushion; back cushion; adjustable height arm rests removable for transfers; quick-release rear wheels for easy breakdown and storage in a vehicle; correct seat height for transfers and propelling yourself forward; and appropriate seat width in order to go through doors at home.

A power wheelchair for someone with ALS usually includes: special electronics which can be expanded as your needs change (e.g, if hands become too weak, hand controls can be changed to chin or head controls; sip and puff device, or switch); reclining back with head support with a separate motor and switch to control positioning (models may have a back recline only feature or a recline and tilt feature which enables the entire seat and back to tilt back like a rocking chair-this is more expensive); and power operated elevated leg rests.

Not only can a chair help you get around, many of the things you need to do in a day can be done from your wheelchair with adaptations. This makes for fewer needed transfers and reduces the risk of injury. For example, trays can be affixed for eating or holding a laptop computer. Electronic switches used to operate the computer or any other environmental controls in your home can also be attached.

The above chairs have been described to alert you to what is ideal. Your health insurance coverage and other available funding will determine what options are available to you. **Whichever type of wheelchair you use, it is critical to have it customized to best fit you and your mobility needs.**

- **Transfers**

Assisted transfers are a leading cause of **caregiver injury** when proper body mechanics are not used. Poor transfer techniques also increase the risk of endangerment for the person being moved. The following transfer strategies can help, but always learn and practice transfers with a therapist.

1. Transfers Without Equipment

Sit-to-stand transfer: When rising from a seated to standing position (whether alone or with help), scoot to the edge of the chair, lean your trunk forward 30-40 degrees and rise. If someone is helping you, they should squat down facing you and grab your belt or under your buttocks (do not pull on helper's neck or back), then block your knee with one of their knees, and then both of you stand at the same time. When standing, the helper should pull your pelvis toward him/her. Helpers should bend at the hips and knees and not the waist.

Standing pivot: After you have been assisted to stand, and now want to turn, your helper should face you, holding your pelvis closely to theirs and pivot 90 degrees without twisting their spine. Making small steps, shifting weight from side to side can assist in the pivot. To sit down on the surface you are being transferred to, your backside should be in front of it. Then your caregiver should keep one of his/her knees in front of your knee and then bend at the hips and knees as they lower you down, ending up in a squat while you end up in a seated position.

“ I wouldn't want to be without satin or sateen sheets-they allow for easier movement and shifting position, enabling a longer period of independence. ”

Sitting transfer: If sit-to-stand transfers become too difficult, sitting transfers can be substituted. To complete, position a chair (without arms) next to the surface you are transferring to. Scoot your bottom to the edge of the chair. Your caregiver can help you shift your body weight to one side and then pulling the other side of your pelvis toward the edge, and so on. While you lean forward, your helper will squat, block on or both of your knees with his/her own, grab your belt at the back or under the buttocks, rock you forward until your bottom lifts, and then swings your bottom toward the surface you are moving to (bed or seat). If this is too difficult, or the caregiver finds it hard not to twist his/her spine, a transfer board should be used.

Lying-to-sitting: Roll to your side facing the edge of the bed you want to sit up on. If you cannot roll yourself, your caregiver can put you in this position. Once you are on your side, bend your knees so your legs drop at the edge of the bed and then push with your elbow and hand and sit up sideways. A caregiver can offer stability for you at any stage of the move you require. To lie down from sitting, you can reverse the maneuver.

Scooting in bed: When you are transferring to your bed to lie down, always try to position your bottom in such that when you recline your head to end up in the correct spot. If you need to scoot yourself around while lying on your back, bend both knees, keep feet on the bed, and lift up your bottom a little. Then you can move your bottom in the desired position. If help

is needed, your caregiver can help to either hold your feet down or use a towel to support your bottom and lift.

2. Transfers with Equipment

Transfer board: A smooth board acts as a bridge between two surfaces and is very helpful when you have trouble standing. With one end of the board under your bottom, and the other sitting on the surface you are transferring to, you slide across the board with caregiver

assistance. This can be difficult if you are transferring to a higher level surface because you are going up hill.

Rotating transfer board: This is a board that has a lazy Susan feature on it that will rotate you once you are positioned on it. This takes practice, but can be very effective. A common brand of rotating transfer board is the Beasy® Board.

There are other transfer devices such as a standing transfer pivot. Ask your therapist for the latest in available equipment.

- **Lifts**

Lifts are valuable assistive devices especially when a smaller person has to move a larger person. A commonly used lift is called the Hoyer lift. Although it looks big and perhaps complicated, it really is not. Talk to an equipment specialist when choosing a sling for a lift. They come in various materials some of which are better for bathing than others and some have commode openings.

Another commonly used lift is the E-Z Pivot Lift which does not use a sling and leaves your bottom clear for clothing removal in order to use the toilet.

There is also a lift that can help you stand from a seated position. These are called Easy Lift Chairs and can be very helpful if you still have the ability to stand. This is an electronic lift inside an upholstered chair. Some models also recline. Make sure it also has arms that rise with the seat to stabilize you.

Terry Schultz
ALS family caregiver

Other types of lifts that are not discussed here are available for installation in pools, tubs, and vans. Ask your therapist for additional information and resources.

TRAVEL TIPS

- **Facilities**

If you have problems walking, or are in a wheelchair, telephone your destination ahead to find out what accessible facilities are available where you will be staying, as well the attractions you may visit. Some places are very accessible to the disabled, others are not. Be very specific about what your needs are - "accessible" means different things to different people. So, you may need to ask if there are ramps, elevators, wheelchair accessible rooms in the hotel, etc.

- **Air Travel**

Most airports provide wheelchairs that you can take to and from the plane. Make sure that the airline knows in advance that you require a wheelchair. If you need to be accompanied by an attendant, and have the appropriate document signed by a doctor, some airlines let an accompanying attendant fly at half-price. Airlines also provide other services for the disabled including special meals. **When inquiring about reservations, it is also a good idea to make sure that the airline can accommodate your disability requirements.**

- **Auto Travel**

Some families with ALS purchase a wheelchair accessible (barrier-free) van, or one with hand controls for persons with good upper body function for every day use as well as road trip vacations. **There are companies that also rent hand control adapted and barrier-free converted vans** for extended road trips, or to use upon arrival at your destination. For the most part these companies are based in the United States (e.g., Wheelchair Getaways, (800) 642-2042, www.wheelchairgetaways.com). For auto travel in Ontario, barrier-free van rentals are available in Toronto through Kino Mobility at (888) 495-4455. Check with your local Canadian Automobile Association (CAA) or destination Visitors Bureau to find out about other resources that may be available in your destination city.

- **Support Resources While Away**

If you are traveling to another part of Canada, make sure you know how to contact the ALS Society office closest to your destination for information on the services they provide in the event you will need them. Similarly, if you are going to be traveling in the United States, contact the nearest ALS Association (ALSA) Chapter. For an ALSA Chapter list, go to www.alsa.org. If you are traveling abroad, check the ALS/MND International Alliance web site for information on associations available by country (www.alsmndalliance.org).

A Final Note on Mobility...

Mobility function and assistive needs will change over time with ALS. It is important to remain in contact with occupational and physiotherapists who understand ALS as they can help you plan for changes and adaptations ahead of time. Advance planning will help make transitions smoother as well as promote comfort, safety, and independence.

Please access the ALS Society nearest you to inquire about an equipment program designed to get you the devices you need in as timely a manner as possible. Together, you, your healthcare team and the society can work to help you adapt to mobility changes and maintain independence for as long as possible.

Some people with ALS may lose function in their mouth and throat which makes eating and drinking a challenge. Due to fear of choking, or frustration and fatigue with the time it takes to eat a meal, some individuals may eat less, resulting in not getting enough nutrition (calories and nutrients), weight loss, worsened fatigue, and weakness.

Fatigue plays a very big role in why a person's nutritional status declines. Weight loss occurs because with ALS, increased energy is required for all activities including eating and your body's basic energy requirements are higher (increased basal metabolic rate).

Therefore, addressing issues with the mechanics of eating and drinking (controlling food and swallowing) and alternative dietary solutions is critical in the management of ALS. The speech language pathologist and dietitian on your healthcare team, along with your doctor, will be your major resources for adapting to swallowing problems and maintaining good nutrition.

THE MECHANICS OF SWALLOWING

Swallowing is the act of moving food or drink from the lips to the **esophagus** (food tube). If your swallowing muscles are affected, your symptoms will depend on which muscles are weak. For example, if you have tongue and lip weakness you may have trouble keeping liquids sealed in your mouth for swallowing or moving food around in your mouth effectively. If your jaw muscles are more affected, chewing may be tiresome or difficult.

The Four Phases of Swallowing

In **phase-one**, the food is chewed or "processed" in the mouth to form a **mass**. In **phase-two**, the muscles of the cheek, tongue, and upper throat move it into the back of the mouth where the food triggers a **reflex-like response causing the soft palate to rise to prevent food from entering the nasal cavity** (nose). If these muscles are weakened, food may be difficult to move back in the mouth, or enter the throat prematurely.

In **phase-three the larynx (voice box) is raised**, and the tongue moves back and the airway entry is closed to prevent food from entering the **trachea (windpipe)**.

In **phase-four** the muscles at the top of the esophagus contract (people with ALS may find that the lower muscles don't relax, in which case food gets stuck, feeling as if it is stuck in the back of the throat). A final muscle contraction moves the food into the esophagus toward the stomach.

Swallowing Assessments

Swallowing assessments typically involve a chair side assessment which involves inspection of your mouth, and observations of how you eat and drink small samples of food and liquid. Sometimes you will be asked to eat or drink different consistencies of food and drink to determine which are easiest for you to swallow. Throughout all assessments, your therapist will be interested in your experience with swallowing and will be asking detailed questions about foods and drinks that cause you to cough or clear your throat.

It is also common for swallowing assessments to include an x-ray procedure called a **Modified Barium Swallow Videofluoroscopy**. This is ordered with the permission of your doctor and is an outpatient exam. You will be asked to eat and drink small amounts of food and drink that will be coated in barium so they can be seen on x-ray. This exam is helpful in identifying swallowing problems and testing "tricks" that may help you swallow more safely.

Because of the variety of muscles used to speak and swallow, the problems that occur as a result of degeneration depend on which muscles have been affected. **If these problems occur it is important that you consult your doctor for a referral to a speech-language pathologist with ALS knowledge and expertise in assessing and managing swallowing problems.** It is sometimes valuable for you and your caregiver to see a copy of your swallowing x-ray to give you a better understanding of what happens inside your throat when you swallow.



Common Swallowing Difficulties

Swallowing difficulties may include:

- Coughing
- Throat clearing after a sip or bite
- Leaking of food from mouth
- Difficulty chewing
- Difficulty moving food around mouth
- Difficulty starting to swallow
- Food or drink escaping from nose area
- Sensation of food caught or stuck in the throat

You may also want to consult a therapist if you:

- Get tired during meals
- Take a long time to eat (longer than 30 minutes per meal)
- Lose weight
- No longer enjoy or are uninterested in eating

Adapting How You Take Medication

Many people with ALS have particular trouble with swallowing medications. If you are having this problem you might consider:

1. Grinding pills into powder with a pill crusher (available from your pharmacy) and mix them with pudding or another smooth, easy-to-eat food like apple sauce or yogurt (ask the pharmacist if it is appropriate to crush pills-it is not recommended for slow release medications)
2. Asking your pharmacist if your pill medication is available in liquid form, or if he or she could custom make a liquid suspension for you.

ADAPTING HOW AND WHAT YOU EAT

How to Make Eating and Drinking Safer

While eating can be one of the joys in life, it can be a challenging activity for people with ALS with weakened bulbar muscles. There is a **well documented connection between safe swallowing and maintaining good respiratory (breathing) health.** People with swallowing problems are at higher risk of developing a pneumonia called aspiration pneumonia than the average population. Learn as much as you can about your own swallowing limitations and make sure your diet is adapted to your changing abili-

ties. Here are some tips for making eating easier and safer.

- **Take more time**

Eating and drinking may be a very slow and labour intensive process. Allow more time to eat meals and avoid rushing.

- **Be relaxed when eating**

A person experiencing difficulty with eating and drinking may feel acute embarrassment. Anxiety and distress may accompany embarrassment, and anxiety itself impairs the ability to relax. Being relaxed and feeling confident is of tremendous assistance.

- **Concentrate on eating**

Make sure there are **no distractions** like TV or radio while eating. If eating in a group is also distracting, consider quieter meal times for yourself and one other person who can help you when needed. You can then simply use the group meal time to socialize and not worry about eating at that time. Do not try to combine eating with talking.

- **Focus on eating position**

With swallowing problems you should eat in an upright position. Sometimes your SLP will suggest you tuck your **chin towards your chest** to further protect your airway during feeding. There is also adapted feeding equipment which your occupational therapist (OT) may recommend for easier eating.

- **Take small bites**

Small bites will require less trouble chewing and managing food in your mouth. Also, if you have problems with liquids, use straws with caution as liquid can be brought up into the mouth at a higher speed and volume than may be safely swallowed.

- **Take food and liquid separately**

It may be difficult for the swallowing muscles to switch between eating and drinking, which require slightly different muscle activity. If you have foods like soups that have broth and solids, it may help to take in one consistency at a time, i.e., take a sip of broth and then a spoonful of solids.

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- **Emergency Preparedness**

Learn what to do before an emergency happens. When the throat gets irritated or blocked, it can close around the irritant making it difficult to breathe. Even your saliva can cause coughing and choking. This may also happen when a few small particles from a previous meal are still stuck in the throat.

It is best that you and your caregivers take a recognized First Aid course with special attention given to identifying signs of obstruction (blockage) and learning appropriate responses.

Knowing the Heimlich maneuver is a must for your caregivers. Ask a member of your ALS care team about this or contact your regional ALS Unit, local Red Cross Society or St. John's Ambulance branch for CPR and First Aid courses in your area.

The emergency response suggestions below may or may not be appropriate for every individual circumstance. Reading these is by no means a substitute for First Aid certification. They are included here just to give you some familiarity with what may be helpful.

- **Try swallowing two or three times to clear it.** It may be that the upper oesophageal sphincter muscles at the bottom of your throat, which are normally closed, are not relaxing and opening as actively as they should to allow swallowing.
- **Try to relax.** Lean forward, the further the better, depending on the seriousness of the problem. If possible, stand up and bend over as if to touch your toes
- **Try to take small breaths through the nose.** Expel the food by breathing in slowly, and exhaling or coughing quickly. For more force "splint" you're abdomen by crossing your arms over your lower abdomen and pressing firmly in and down as you cough out. A caregiver can also do this by standing behind you and pressing their hands into your abdomen as you cough. Ask a nurse or other professional to show you this technique, known as the Heimlich maneuver, and practice it with your caregivers as soon as possible so that you will know what to do should choking occur.

Things not to do in an emergency:

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- **Never use liquid to wash down food** If food is stuck in the throat and liquid is added, the liquid can very easily be channeled into the airway leading to your chest, and cause choking.
- **Never hit a choking person on the back** When a person is choking from food, hitting on the back can cause the food to jam tighter in the throat.

Dietary Changes that Can Help

If chewing and swallowing problems develop and progress, it may be necessary to make **dietary texture changes** to cope with these reduced eating abilities. Consult with your dietitian, swallowing therapist, or doctor regarding changes to your diet. With changes in your oral skills (use of tongue and lips), it may not only be difficult to chew, but also difficult to control food in your mouth. You may find it hard to keep your lips sealed tightly during eating or drinking. Or some foods and liquids may "get away from you" before you are ready to swallow them.

When eating or drinking consider the following:

- **Choose smaller and softer foods**

Food should be softer and cut into smaller pieces that require less chewing.

- **Avoid very thin, runny foods**

If food or drinks are too thin or runny, they may be harder to control in the mouth and escape into the airway before you are prepared to swallow them. Some of the liquid can run into the airway to the lungs and cause coughing.

- **Avoid very dry foods**

Crumbly, dry or flaky foods like day old muffins, toast, crackers, chips, pastries, dry fish, and dry mashed potatoes may be more difficult to manage due to their tendency to have loose and errant crumbs. Moistening solids with gravies, sauces, butter, and broths can make dry foods more manageable.



- **Avoid mixed consistencies**

Some people find that foods that have thin liquids mixed in with solids, like cereal and milk, or soup broth with meat or vegetables problematic because they have to manage two different food textures at the same time. Try taking either the liquid or solid separately. The same could be said for skinned solids, like peas, corn or oranges, which have both a skin and soft or juicy centre.

A dietitian can make helpful suggestions about the following:

- Suitable methods of cooking
- Substitutions for suitable foods
- Ways to thicken thin liquids (for example, use commercial thickeners, or natural thickeners)
- Semi-solid foods (e.g., pudding, custard, cottage cheese, puréed fruit)

MAINTAINING GOOD NUTRITION

It is challenging to maintain a fully balanced diet if you have chewing and swallowing difficulties.

However, it is so important to maintain good nutrition to enhance your strength and energy levels.

Your nutritional needs remain high, even though your activity level is low.

There are many ways to add nutrients to food - this is not the time to worry about fat and cholesterol! In addition to special food supplements available in stores, there are many natural ways to supplement your food and drink for increased nutritional value. **Please consult with your registered dietitian for appropriate ways to maximize your nutrition.** Drinking enough non-caffeinated liquids is extremely important for overall health as well as for energy.

If you haven't already done so, now is the time to discuss with your doctor the option of having a feeding tube put in place to make sure you maintain good nutrition.

Tube Feeding

- **What's it all about?**

Getting a feeding tube allows you to take in food and drink for energy and nutrition without having to do so by mouth. When swallowing and chewing are very difficult, you will not be able to get enough nutrition and hydration by mouth alone.

If you decide to have a feeding tube, the recommendation is "the earlier, the better-even before you need it." The procedure is easier to tolerate and it gives you a chance to get comfortable with it before you actually have to use it.

SWALLOWING PROBLEMS: FOODS TO LOSE...

- Extra-spicy, "hot" foods
- Acidic foods like pickles and some dressings
- Soft, doughy fresh bread
- Cookies, crackers, dry cereal, graham crackers
- Dry muffins, cake
- Dry, fibrous, or bony meats and fish
- Fruits with tough skins
- Stringy fruits (e.g., pineapple, oranges, watermelon)
- Stringy vegetables (e.g., lettuce, celery, string beans)
- Fruits and vegetables with skin or seeds (e.g., peas, corn, apples, berries)
- Fried noodles, rice
- Popcorn, potato chips, nuts
- Foods with small particles (e.g., nuts, seeds, coconut)
- Sticky foods (e.g., peanut butter)

SWALLOWING SOLUTIONS: FOODS TO CHOOSE...

- Custards, apple sauce, sherbets, puddings, plain yogurt
- Moist meats with gravy, sauce, or broths to moisten
- Canned fruit, soft fruit (e.g., bananas)
- Eggs (scrambled, poached, NOT fried, and omelettes)
- Cooked cereals (with milk)
- Casseroles (macaroni and cheese)
- Milkshakes, frozen ice cream bars
- Mashed potatoes with gravy
- Gelatin with yogurt/cottage cheese
- Salmon/tuna/egg salad moistened with mayonnaise or other spread
- Thick creamed or puréed soups
- Juice nectars, tomato juice, vegetable juice
- Pasta with sauce



Some people may not require using it immediately (although it will still need daily flushes of water); some may use it to supplement intake in addition to eating; and some may switch from eating to total tube feeding. **You and your dietitian should discuss which scenario fits you best and together develop a plan to meet your nutritional needs and goals.**

The following information is designed to give you a better idea of what getting and living with a feeding tube is all about and to help you with your decision-making process.

- **The procedure**

If you make the decision to have a feeding tube, your doctor will refer you to a **gastroenterologist or a gastrointestinal (GI) or general surgeon.**

To insert a stomach feeding tube, commonly referred to as a **percutaneous endoscopic gastrostomy (PEG)** tube, requires a simple 30-minute operation under mild sedation. A gastroenterologist will perform the operation. The tube is placed directly into your stomach through the abdominal wall to provide another way for you to receive nourishment and liquids.

Although the most common feeding tube for ALS patients is the PEG, your doctor may recommend getting a gastro-jejunostomy feeding tube (GJ-tube) instead. The procedure and breathing function requirements differ. Talk to your doctor about your options.

There are different types of feeding tube materials. Discuss which type is best for you with your doctor. PEG tubes can be either an actual tube made of soft pliable material which extends about six inches outside of your stomach, or in a button form which is smaller and lies flush against your abdomen. The feeding tube remains in place and is capped and uncapped at each feeding. Initially, a dressing is required around the incision. A small amount of leakage around the incision is not unusual. When the incision heals, leaving it open to the air may be the best way to keep it healthy.

- **Post-operative adjustment period**

After the operation, you may have to stay in the hospital so that healthcare professionals can assess your body's ability to adjust to an alternate way of getting nutrition. How long a patient stays is dependent on his or her health. In some regions, the procedure may be performed as out-patient surgery followed by immediate home care support. Care providers will prescribe the quantity and type of nutritional supplements you should receive through tube feedings. During this period, learn how your new tube-feeding system works and how to manage your tube feeds. Even if you won't be using tube feeding right away, you will still need to practise flushing the tube twice per day to keep it clean. It is also important for your family or caregivers to receive the same training.

- **The feeding pump**

Tube feeding can be accomplished through what is called "gravity feeding" by hanging the feed bag from an IV pole without a pump. However, using a feeding pump for assistance may be more comfortable and less likely to result in gastrointestinal problems (fullness, gas, regurgitation, vomiting, and diarrhea).

Portable pumps are available for greater mobility. Talk to your dietitian about feeding pumps and other supplies.

- **Excessive coughing may occur**

You may find that tube feeding causes excessive coughing. This may happen for a number of reasons, including excess saliva, not sitting up enough, the feeding rate is too fast, or because of various other stomach problems.

If this happens to you, consult your dietitian who may suggest reducing your feeding rate and/or decrease the amount of formula given at the feeding.

- **Avoid lying down during and after feeding**

You need to remain upright during the feeding and for another 45 minutes after the feed is completed.



• Ask for home care tips

After you have a feeding tube installed, and before leaving the hospital, arrange for appropriate home care assistance while you are getting used to tube feeding. You will probably have some questions regarding tube feeding, cleaning procedures, supplies, suppliers, government assistance programs, etc. Also, hooking up the feeding bag requires a few tricks to prevent sending a tube full of air into your stomach before the food.

• Avoiding dehydration

It is a serious mistake to assume that you are getting enough liquid because you are living on liquid nutrients. Concentrated liquid nutrients do not contain much water. Take lots of water to avoid dehydration. Your dietitian will include in your tube feeding schedule how much water you need.

• Choosing Tube-Feeding Equipment

When you first have your feeding tube installed, you can learn in the hospital what your equipment needs will be. When you are at home you may first be using equipment supplied by your local ALS Society or home care provider for a few weeks, after which you will need to get your own equipment. You will need a method to hold up the container while feeding, such as an IV pole, a supply of feeding containers, some feeding syringes, and a supply of liquid food.

• Tube-feeding diet

A dietitian at the hospital will usually work out your tube-feeding requirements based on what you can

still eat normally and your estimated calorie requirements. Because your eating abilities will change, your tube-feeding requirements will probably increase over time. When this occurs, increased feeding tube requirements should be recalculated by a dietitian. Follow-up may also be provided by a home care or community dietitian.

If you are not a patient at an ALS clinic, it is important that your doctor and dietitian be educated about ALS. Encourage them to contact your local ALS Society for referrals to specialists with whom they can consult.

WHEN TO CONSIDER A TUBE...

- When your weight goes 10% to 15% below normal, it is a good time to consider getting a feeding tube. You need to ask your doctor what a "normal" weight is for you.
- When eating or drinking leads to frequent choking
- When it takes more than one hour to eat a meal
- When eating is no longer a pleasurable activity
- When respiratory function is decreasing and risk of aspiration pneumonia is increasing

WHEN NOT TO CONSIDER A TUBE...

- When your breathing function is considered too poor (see *Adapting to Changes in Breathing and Maintaining Lung Function* sub-section)
- When the idea of an incision or tube in the abdomen is unacceptable
- When the cost of the formula is too high
- When the risks outweigh the benefits

You should discuss the various commercially prepared alternatives with a dietitian. Depending on where you live, products available may vary. However, some of the commonly recommended products are Boost®, Jevity®, Jevity Plus®, Nutren®, and Nutren® with Fibre, Isosource®, Resource®, and Compleat®. Your dietitian will prescribe what is best for you.

• Making your decision about a feeding tube

Making the decision to have a feeding tube is a **major care decision when you have ALS**. Some people choose to have a feeding tube, while others do not. Each deci-

sion should be made based on available, accurate information and personal preferences. Whatever you decide, you should **document your choice in your personal health care directive** (Living Will-see *Section 6 Legal and Financial Considerations*), share a copy of your directive with your doctor, and discuss your decision with your family members, in particular the individual whom you will appoint to

be your proxy (legal representative of your wishes) in the event you cannot communicate your wishes to healthcare providers. Use the table on page 42 to review issues to consider when making a feeding tube decision.

A Final Note on Swallowing and Nutrition...

If you have changes in swallowing, you may need to make gradual changes in how and what you eat and drink. Working closely with your doctor, dietitian, and speech language pathologist, will help you manage your nutritional challenges while maximizing the enjoyment of tasting and eating for as long as you can.

Eventually, you may need to make a major decision about whether or not you want to have a feeding tube. Use the information provided in this publication as a discussion tool for you and your doctor and family members early so your decision is one that is thoughtful, timely, and right for you.

If you experience weakening muscles of the face, throat, neck and tongue, you may find difficulty with speaking, chewing, swallowing and controlling mucous and saliva. If the onset of ALS is associated with these types of symptoms, it is known as "Bulbar ALS." People first diagnosed with limb onset ALS may develop bulbar symptoms later on.

If speech problems occur, they progress gradually. Therefore, **it is important to work with a speech language pathologist (SLP) with experience in ALS and augmentative and alternative communication (AAC) early on, even before there are any speech impairments** to assess speech, monitor it over time, and teach you strategies to help keep natural speech for as long as possible, while at the same time gradually introducing other means of communication. Adaptations can and should be made to ensure you can always communicate what you want, when you want.

CHANGES IN SPEECH

A speech language pathologist's assessment is helpful to determine which muscles are weak and how to best use the muscles still available for speaking. **Speaking problems are generally caused by weaknesses in one or more of the following muscle groups:**

- Breathing muscles - resulting in less air flow available to power your voice resulting in a strained, strangled sound;
- Vocal cords - resulting in breathiness, lowered pitch, and/or monotonous sounds;
- Soft palate and throat muscles - resulting in a nasal sound;
- Tongue and lip muscles - resulting in difficulty forming word sounds.

The following symptoms may be experienced:

- Weakness, stiffness, slow movements in your mouth, throat, and jaw muscles
- Muscle atrophy (wasting) or fasciculations (twitching) in your mouth and throat muscles
- Changes in voice quality (hoarseness, strained-strangled, lower pitch, nasal, breathy, monotone, volume changes)
- Changes in your speech (nasal sounds, slower speech, not precise)
- Difficulty making your speech understood

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- Increased difficulty with speech as the day goes on (fatigue)

Losing the ability to speak is a very significant change and one of the most difficult challenges associated with ALS. It can be frustrating for both the communicator and the listener. For some, it may be the end of off-the-cuff remarks and participating in quick conversation. For the listener it may become a question of trying to interpret. Some people are much better at it than others.

It is important to openly discuss how to adapt to changing speech with communication partners to maintain the best possible connection to family, friends, and caregivers. Caregivers need to make adjustments as well such as giving the person with ALS extra time to speak, encourage and accept different forms of communication, and problem solve together when communication breaks down.

It is important to remember that loss of speech is gradual and there is help. A range of strategies to enhance communication should be explored between you, your caregivers, and SLP. While adapting speech patterns when speech is still the main way of communicating can help, using alternative communication methods, as described more fully in the next section, will need to be considered if the ability to speak is substantially lost, or if it makes it easier to communicate in some situations even when you have some speech ability.

Use of an **oral prosthesis** (palatal lift or other hard-palate prosthetic device) may be considered as a **short-term option** in cases where existing speech can be improved. Typically a palatal lift reduces loss of air through the nose (nasal speech) and increases air pressure required for sound production. It cannot compensate for weak lips, breathing, or vocal cords. Sometimes a prosthesis can be added to the palatal lift to lower the hard palate of the mouth. This allows the weakened tongue to make contact with a hard surface to produce sounds such as "t,d,k,g".

A prosthesis may improve swallowing and reduce problems associated with excessive salivation. However, it may not be a realistic option in cases where there is rapid change in communication function and control, or a strong gag reflex. You should consult a

speech language pathologist and/or prosthodontist (dentist who specializes in oral devices) to determine your best option.

Use of a voice **amplifier device** can enhance the volume of the voice. They may be an option if speech is understandable, but respiratory weakness is the cause of decreased speaking volume.

Strategies to use later as speaking muscles continue to weaken may include the following:

- Use words that are easier to pronounce, if forming words is a problem
- Carry a pad of paper and pen to write out your messages if you can still use your fingers
- Work out hand signals for frequently used phrases with your immediate family and other caregivers
- Use non-verbal signals (e.g., eye blinks for yes or no)

Changing your patterns of speech can be difficult to do and may require practice sessions with an SLP to make those changes.

AUGMENTATIVE AND ALTERNATIVE COMMUNICATION (AAC) STRATEGIES

AAC refers to a system used to either augment (enhance) existing speech, or to serve as an alternative when there is no speech. AAC does NOT only mean using a "computer talking device." AAC strategies refer to a range of methods used to facilitate sharing information, self-expression, signaling for attention, maintaining contact with others, and problem solving.

Addressing AAC issues early will enable you to feel better prepared and comfortable when changes occur. If high tech interventions that record speech are going to be considered, you can record your voice for future use in such devices. This may be particularly preferable when you have young children.

FACTORS IN MAKING AAC DECISIONS

Think through what your needs are, and will be in the future. For example:

- Will you need a device mostly for communicating commonly used phrases?
- Are you able to write messages?
- Will you have to make presentations to groups of people?

When choosing a device, some of the following factors may be relevant:

- Level of training needed and the complexity of the system
- Cost/coverage
- Portability
- Versatility
- Speed of communication
- Circumstances of use
- Amount of follow-up needed

Which AAC strategies are best for you depends on your functional ability and on the situation.

You may end up using several different ways to communicate over the course of a day. For instance you may use speech to communicate with your family members who are used to hearing you and can see you. E-mail may be preferred to speaking on the phone with others at a distance. You may write messages or spell using a letter board or use a device that talks for you when communicating with people who are less familiar with your speech.

Some people want to avoid electronic AAC devices and use only low-tech strategies and tools while others want the latest, most sophisticated high-tech equipment available. Each device should be chosen with the unique needs of the individual in mind. You should also be aware of how long the chosen system will be useful to you. It is most likely that you will need to use several strategies as the disease progresses.

People using invasive ventilation must contend with a tracheostomy tube inserted into their throat. But, they may be able to speak by using a speaking valve (e.g., Passy-Muir). A respiratory therapist can help train how to use this device.

Before purchasing any AAC devices, get advice from an assistive technology clinic, from an SLP, or from another qualified source such as a rehabilitative engineer, or assistive technology professional. Expert professionals can help you select devices and train you how to use them. You can also visit local suppliers and try out the various options once an SLP or assistive device specialist has made some recommendations. Check to see what devices your local ALS Society has available for you to try.

No-Tech Strategies

Speech clarification and communication partner adjustment strategies require no technology. **Strategies to use when speaking muscles start to weaken** are:

- Conserve energy
- Take in a full breath before speaking
- Speak slowly with short sentences, and words with fewer syllables
- Say each syllable clearly (over exaggerate) and put pauses in between words
- Identify the topic first before you go into further detail
- Make your environment as communication friendly as possible: reduce background noise (e.g., turn TV off or on mute), make sure you and your listener can see each other
- If able, use gestures to what you are saying to add extra information for the listener

“I know how frustrating this is [unable to speak], but I have learned to live with it. My best friend is my FAX MACHINE. I can keep in touch with my friends in Canada, America, and England. The minute I want to say something, I write it down and then feed it into my friend and it is immediately transferred to wherever I wish it to go!”

Bobbi Harris, Age 84
Calgary, AB 2004

must be. At the end of this sub-section, there is an example of a communication board that incorporates all three: letters, topics, and frequently used messages (Beuleman and Miranda, 1998). You simply point to the frequently used message, or letters to spell words.

If you can no longer point to letters, then your eye gaze can be a reliable method of communicating. Communication boards that use gaze are transparent (see-through) and usually feature alphabet letters, symbols and/or complete words, phrases, or sentences. To use the board, look at the desired message or combination of letters.

When only face-to-face communication is needed and you can still indicate selections on a board by pointing or eye movements, a word or letter board is a good

low-tech communication choice.

An SLP can help develop and modify communication boards as needed. You may have several different boards for different communication scenarios. For example, one board may be dedicated for daily care routines, while another is used for TV watching.

• Signaling Systems

These no-cost systems are usually worked out between you and your regular communication partners. **Signaling makes use of facial expressions, eye contact, eye movements, gestures, touch and body language.** Some of this system's drawbacks include the limited nature of possible responses and the inability to communicate with people who are unfamiliar with the system. Personal signaling systems are a valuable backup to high-tech systems that are vulnerable to system failure.

A 24-hour monitoring alarm (for example, a bell, intercom, or buzzer) is necessary for you to get attention when your communication partners are not in view.

You and your family may want to apply for an **emergency response service** especially if there are times

Low-Tech Strategies

Often simple manual systems are preferred because they permit continuing human contact without requiring the higher levels of energy, technical knowledge and skill and motivation needed to operate more sophisticated equipment.

• Writing Boards

Writing boards such as **white boards and magic slates** are often the preferred choice when you can still write. Markers and pens can be built up by an occupational therapist as your grip weakens. Once grip is too weak to hold a marker, you may choose to use a communication board.

• Communication Boards

Communication boards may include **frequently used messages, topics, and/or letters.** Words usually included on communication boards are lists of foods, comfort items, positioning requests, and social phrases. The more information there is, the larger the board

you will be in your home alone. Those who join are given a small (about the size of a small box of matches) transmitter with an emergency button on it. When joining an emergency response system you must tell the program people:

- Facts about their illness
- The names and phone numbers of people who should be contacted in case of emergency

When the emergency button on the small transmitter is pressed, a signal is sent to the service that tells them who has the emergency. They will know the client's illness and immediately call the names given to them for emergency purposes. Ask your healthcare provider or contact the ALS Society in your area for referrals to emergency-system vendors in your area.

- **Manual Communication Aids**

Assistive devices to aid manual communication (i.e., writing, typing, or pointing to communication boards) include **pencil grips, magic slates, and other portable boards, book holders, tilt-top tables, pointers, typing sticks (held in the mouth, hand, or foot), writer's splints, TTY (teletyper), TTD (telephone for the deaf), and page turners.** If you leave home without a caregiver, it may be wise to wear a Medic Alert (medical emergency) bracelet.

- **Small Digitized Communication Devices**

These devices are useful to use for occasions when you need specific **pre-programmed messages.** All models use digitized speech, similar to answering machines and voice mail services. They are a more affordable option to high tech devices described below, however they do not offer the same flexibility as the high tech devices. These low tech devices may offer as few as one to four, and as many as 32 messages on one display.

Some devices come with six, eight, or 12 "levels". That is, a unique, different message display is created for each level and each level includes a different set of messages. This is useful for people who need more than eight or 32 messages in total, but cannot cope with any more than eight or 32 at one time. You simply switch to a new display representing a different conversational topic, change the level on the device by turning

a knob, and you now have more messages available.

- **Specialized Telephone Equipment**

Special phone services are available. If you are unable to lift a phone receiver and/or dial a phone number, there is a **hands-free phone, activated by blowing on a switch, plus an operator dialing service.** Speaker phones may also be helpful. Ask an occupational therapist for more information about what is available in your area.

High-Tech Strategies

If you are interested in a high-tech communication system, information about systems that meet your preferences, your needs and functional abilities should be discussed with an SLP. These systems are very expensive and there is a wide range of high tech devices available. They vary in size, function, application, and operation. Some function by text-to-speech; that is you simply type your message and the device speaks it for you. Some will store commonly used messages, phrases, and/or words. You choose the messages that you wish to store with the assistance of the SLP. To operate, you select the combinations you want and it speaks for you. Many systems offer text-to-speech and pre-stored messages.

Rate enhancement features appear on most of these high tech devices. The three most common **rate enhancement features** include word prediction, abbreviation expansion, and a menu of the most commonly used 100 or so words.

- **Word prediction:** special software loaded in the system predicts the next word after one or two letter selections based on spelling and/or grammatical rules. Typically three to eight possible words are displayed for selection. The options change each time a new letter is entered. Systems that also predict words based on grammar offer word options before you even begin to spell the next word. Some systems even learn the words that are used most often. This makes communication much faster.
- **Abbreviation-expansion:** allows the person using a device to use short cuts to speak longer messages. A common one we use in writing is "ASAP" meaning "as soon as possible". Again this makes communication much faster.

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- **Commonly used words:** These words are always present on the system for the person to use instead of spelling them. It is much faster to select a frequently used word (for example the five "W" questions-Who? What? Where? Why? When?) than to spell it out letter by letter.

Most of these systems can accommodate for changing physical abilities. In the beginning the person with ALS can operate the devices with his/or her hands. The devices can be programmed to accept the lightest of touch should the hands become weaker. Later, if that is no longer an option, the device can be operated by a switch or laser beam using head movement, eye movement, eye-blinks, or any other part of the body that can potentially operate the device.

Many of these systems can be linked to household devices so that the person with ALS can independently control the television, lights, telephone, etc. High tech devices can be classified as follows:

- **Dedicated Communication Devices**

These devices are stand alone communication devices; that is they are designed specifically for communication. They tend to be rugged and travel well. They produce a synthesized (artificial) speech. Some offer both synthesized and digitized speech (recorded messages). They can be used on the telephone, with a group of people, or in quiet face to face conversations.

- **Computers**

Computers can be used for both face to face communication and for written communication. Special software is loaded on the computer to provide speech output and to accommodate for changing physical abilities. They are particularly useful for people who continue to work and are more flexible than a dedicated communication device because they function as both a computer and a communication tool.

Computers are less portable than a device designed specifically for communication. Even laptops tend to be less portable because they are bulkier than a dedicated communication device. Additionally they are not as rugged and thus do not travel as well over bumps and in Canadian weather.

Final Note on Communication...

Although changes in speech and communication function can occur with ALS, when and how your functional speech will change cannot be predicted specifically. However, it is important to know what to look for and how to adapt to changes that happen. Remaining communicative will have a large impact on your quality of life.

Make sure you openly discuss communication issues and adaptive strategies with members of your healthcare team, especially the SLP, **before you have lost substantial speech function.** An SLP or assistive technology expert with expertise in AAC assessments and training will be best able to help you make decisions about what AAC strategies are right for you and your communication partners. Addressing AAC issues early will enable you to feel better prepared and comfortable when changes occur.

Small Talk	Sports	Shopping	Weather
Family	Food	Church	Computers
Personal Care	A B C D E	Yes	Please repeat each word I say so I know you understand.
Transportation	F G H I J	No	You misunderstood.
Trips	K L M N O	Maybe	Start over.
	P Q R S T	I don't know.	This is important.
Appointments	U V W X Y	Forget it.	Wait a minute!
	Z	I have something to say.	
	I will spell the word.		
	I will say the word.		

If you are living with ALS, you may or may not have started to notice some changes in your breathing. While ALS does not impair the lungs themselves, the muscles involved in breathing and coughing will become affected over time. Breathing may be significantly impaired without complaints of breathlessness due to the reduction of physically demanding activities, or wheel chair use. Impairments may not be detected until there is a lung infection likely occurring because of limited breathing ability.

Poor breathing function leaves a person with ALS fatigued, short of breath, and more prone to respiratory infections and respiratory failure. **Therefore, it is a vital part of the ALS disease management plan to monitor breathing function throughout the course of the disease.** However, the reality is that how and when to address respiratory issues in ALS remains debatable and is practised differently across the world. Methods used to monitor lung function vary among doctors even in the same country, or city. Nonetheless, most ALS clinical specialists believe regular monitoring promotes early detection of breathing problems, prevention of unnecessary emergencies, and opens the door for making decisions about therapeutic options available to you in a timely manner.

This publication will explain the breathing process and provide overviews of symptoms of changes in breathing, tests that may be used to monitor breathing function, respiratory infections, airway management techniques, and options for mechanical ventilation and the decision-making process.

THE MECHANICS OF BREATHING

Normal "easy" breathing involves two main muscle groups. When you breathe in, the **diaphragm** moves down; at the same time the **intercostal muscles** between your ribs contract to pull your rib cage up and out. These two actions cause a partial vacuum. Fresh air rushes down the **trachea** (windpipe) through the **bronchi**, the largest air passages in your lungs, and then into the small air sacs (**alveoli**) which pass fresh **oxygen** (O₂) to the blood. This process is called **inspiration**.

When you breathe out, both diaphragm and intercostal muscles relax, and the lung recoils decreasing the size of your chest cavity. Used air that contains waste **car-**

bon dioxide (CO₂) is pushed out of your lungs. This process is called **expiration**.

If you are breathing heavily, two additional muscle groups come into play: when you take a deep breath in, muscles in the neck that attach to the collarbone and upper ribs assist in breathing; and when you force a breath out, your abdominal muscles help to push up the diaphragm.

You can see that there is a great deal of voluntary muscle involvement in the act of breathing. Since ALS is a disease that causes muscle atrophy (wasting) and weakness, it is easy to understand that changes in breathing will occur, even though your lung tissue may be very healthy. Individuals with pre-existing lung disease may be more affected.

Signs and Symptoms of Weakened Breathing Muscles

- **Shortness of Breath with Activity**

Shortness of breath may be the first symptom of the weakening of breathing muscles. You may notice shortness of breath after rushing upstairs, carrying a load, etc. Shortness of breath can also occur with no particular exertion; you may not be able to walk or talk for long without becoming winded. You should tell your doctor about these symptoms. When you experience shortness of breath, stop what you are doing. Shortness of breath will disappear when activity is reduced. Relax in one of the following positions if you are able and breathe slowly and deeply.

- Sit at a table, lean forward with a straight back and rest your head and shoulders on a pillow on the table.
- Sit on a chair or the edge of your bed, lean forward, keep your back straight and rest your elbows and forearms on your thighs.
- Lean against a wall with your feet about 12 inches from the wall. Rest your lower back against the wall and lean your upper back away from the wall.

- **Fatigue**

Fatigue, or tiredness, is a common symptom of ALS. It is caused by a number of factors. As ALS attacks your motor neurons, they become unable to send commands from your brain to the muscle cells that they

control. A smaller number of muscle cells must then try to perform jobs usually done by the full number. The result is that your muscles tire before they normally would.

When your respiratory muscles are affected by ALS, you may be less able to clear the amount of CO₂ you need to and therefore O₂ may also be reduced. When activity increases, it becomes more difficult for the lungs to supply enough oxygen to the body. Other metabolic changes take place and you feel fatigued.

Besides the physiological changes that may cause fatigue, ALS produces many changes in your life. Change often results in general stress that can also manifest itself as fatigue. Do what you can to prevent getting tired. Rest when you begin to feel tired. Try to keep your exertions within the limits that your body is now imposing. Pay attention to signals like fatigue. It may be necessary for you to change your priorities and forego less important activities. Plan your day to include regular rest periods.

- **Morning Fatigue**

Some people with ALS experience morning fatigue. You may wake up feeling tired, sometimes with a headache, and the feeling that you slept poorly. This may be caused by obstructive sleep apnea and/or under-ventilation.

Obstructive sleep apnea: Weakened (bulbar) muscles of the upper throat and pharynx may result in noisy breathing, snoring, or closing off the upper airway while sleeping. This is what is called sleep apnea and is characterized by intermittent reduction or stopping of breathing. A decrease in the oxygen (O₂) saturation of the blood can result.

Hypoventilation: The relaxation of nerve and muscle functions during sleep may lead to under-ventilation (hypoventilation) causing carbon dioxide (CO₂) levels to rise. For example, when you stand upright, the diaphragm moves down when you breathe in. When you lie down, the organs in your abdomen press against the diaphragm, and more strength is required for the diaphragm to move down during breathing.

To help cope with this problem, you may want to try to raise your head and shoulders during sleep. It is

best to raise the whole torso by using blocks under bed 4" to 6" high, a wedge pillow 6-8" at its highest point, or a mechanical bed. If you do not have access to the best solutions, try putting two or more pillows under your head and shoulders until you do.

Obstructive sleep apnea and under-ventilation often occur together, therefore, **nocturnal oximetry** or a **sleep study** may be advised (see **Monitoring Your Breathing Function**).

- **Excess Mucus and Secretions**

In some people, excess mucus and secretions can build up to the point where they cannot be easily cleared by coughing. This is particularly the case first thing in the morning when secretions have built up during the night. If you experience this problem, ask your doctor for a referral to a physiotherapist or respiratory therapist to instruct you in methods to loosen secretions and bring them to the mouth to be spit out.

One method of dealing with mucus and saliva build-up is suctioning. Suction equipment acts very much like a dentist's suction tube; it consists of a motor, collection bottle, tubing and catheters. However, some healthcare professionals believe suctioning may be counter-productive for persons with ALS.

Talk to your respiratory therapist or respirologist about whether suctioning may be right for you, and if there is anything else you can do to reduce mucous build-up, such as assisted cough techniques (see more later in this publication) or use of medications such as Amitryptilline, Transderm V, Atropine, Glycopyrolate, and beta blockers.

- **Weak Cough**

During a cough, the diaphragm and additional muscles completely fill the lungs. Then, abdominal muscles and intercostal muscles between the ribs contract quickly generating a high pressure against a glottis (larynx) that is closed by force. The glottis is then suddenly opened and a very high velocity flow of air is pushed from the lungs and up the airway. Any mucus or food in one of the air passages is forced out as well.

People with ALS have normal cough reflexes, but the muscles involved may be weakened and unable

to produce a strong enough cough. You can strengthen your cough using techniques mentioned later in this section and described in more detail on the web site of The Rehabilitation Centre in Ottawa at <http://www.rehab.on.ca/program/respiratory/neuromuscular.html>. You will need to consult with a skilled professional to train you and your family.

Quick List of Signs and Symptoms of Breathing Problems

- Paleness
- Bluish colour to finger tips (cyanosis)
- Contraction of neck and other muscles to breathe
- Confusion
- Inappropriate sleepiness
- Inadequate cough

MONITORING YOUR BREATHING FUNCTION

There are a variety of methods to choose from to monitor the breathing function of a person with ALS. Physicians vary in their approach, so keep that in mind as you read this section. If you have not had your breathing function monitored, use this educational tool to open a dialogue with your doctor.

Since ALS is a progressive disease, it is helpful to assess pulmonary function early on in the disease, so that there is a **baseline pulmonary function test (PFT)** result, or starting point, to which follow-up assessments every few months can be compared. Without monitoring, a person with ALS could seemingly experience sudden respiratory failure without having had the benefit of advance planning regarding ventilation options. Plus, with use of appropriate airway management strategies, quality of life can be so much better. Signs and symptoms of weakness in the breathing muscles can be subtle and missed if not specifically assessed.

Repeating PFTs every three to six months may be

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advised to assess for any decline in the respiratory system to guide your doctor about when to discuss certain interventions such as airway management techniques, or noninvasive ventilation in as timely a manner as possible.

The following are some of the measurements used to assess lung function.

Forced Vital Capacity (FVC)

FVC is the **volume** of air that can be maximally, forcefully exhaled following maximal inspiratory effort. The test generally involves having you sit with your trunk elevated to between 30 to 90 degrees, putting a mouthpiece in your mouth, a nose clip on your nose, and breathing into a **spirometer** that may either be a simple hand held unit or a computerized machine. After taking as deep a breath as you can, you blow out quickly and as hard as you can for one-to-two seconds. If your lips are weak, you will be provided a lip seal mouthpiece or mask to promote an accurate measurement.

Peak Expiratory Flow Rate (PEFR)

PEFR is the highest rate of expiratory (out) **airflow** you can generate (**peak expiratory flow**) following maximal inspiratory (in) effort. Flow rate can be measured at the same time as FVC using spirometry with flow volume loops. **This is closely related to Peak Cough Flow (PCF)** which tends to be slightly greater. PCF can also be measured at home using an inexpensive hand held **peak flow meter** commonly used by people with asthma.

Normal PCF values would be 360 to 720 Litres per minute (L/min). The minimum effective PCF is 180 to 200 L/min (recognizing that some smaller individuals may manage with lower values). PCF is highly dependent on the function of those muscles in the mouth, throat and larynx (bulbar muscles/bulbar function). The greater the bulbar impairment is, the lower these measures become.

Maximum Inspiratory Force (MIF)/ Maximum Expiratory Force (MEF)

Maximum inspiratory force (MIF) or maximum inspiratory pressure (MIP) is the greatest force gen-

erated by the muscles of inspiration (breathing in), primarily the diaphragm, but also other accessory muscles. A pressure measuring device (manometer) is attached to a mouth-piece and the individual is instructed to breathe in as forcefully as possible without using the cheeks (the cheek muscles can falsely indicate a much higher pressure).

Maximum expiratory force (MEF) or maximum expiratory pressure (MEP) is the greatest force generated by the muscles of expiration (breathing out), primarily the abdominal muscles. These too may be much weakened in ALS. These muscles and their pressure measurement are important in the prediction of an adequate cough. Remember, an adequate cough helps to clear airways.

Maximum pressures of less than 40 - 50 cm H₂O are of some concern as they may be a sign of less respiratory reserve and some limitation of ventilation or cough ability.

Arterial Blood Oxygen Saturation (SaO₂)

A noninvasive way to measure oxygen saturation is to use an **oximeter** device which involves attaching small sensors to a pulse point on a person's body (e.g., finger tip) to transmit oxygen saturation readings. Normal levels of oxygen saturation are above 95%. Lower than expected levels of oxygen saturation in the blood (**SaO₂**) may indicate areas of loss of volume in the lungs (**atelectasis**) or when associated with illness may indicate pneumonia. Less commonly a low saturation can indicate that not enough CO₂ is being cleared (**hypoventilation**). Hypoventilation often begins, or is worse during sleep.

Nocturnal oximetry, is an over-night assessment used as an alternative to a full sleep study (**polysomnogram**) when problems associated with night-time (nocturnal) apneas (short periods of time when breathing stops), or hypoventilation are evident. Sleep studies, conducted over-night in a sleep laboratory, are not commonly performed in ALS management, but in some cases of suspected obstructive sleep apnea and nocturnal hypoventilation they may be advised.

Note: The Relationship Between Monitoring Breathing and Feeding Tube Placement

Another reason respiratory monitoring is important is related to percutaneous endoscopic gastrostomy (PEG) feeding tube placement. About 50% of people with ALS choose to have PEG tubes. What may be over-looked is that **a minimal degree of respiratory function is necessary in order to under-go the procedure in a safe, uncomplicated manner.** It has been recommended that PEG tubes be seriously considered at an FVC of about 50%. Some individuals may continue to swallow very safely, but have a progressive and significant decline in their respiratory function. This means that some individuals who are not at significant risk for aspiration may still need a PEG tube earlier than they would need for swallowing alone because the safe placement depends on their breathing function. In such cases, the tube can remain unused (aside from periodic flushing) until required from the perspective of safe swallowing or nutritional supplementation. Medications that patients may have difficulty swallowing may be given through the tube even if not needed for nutrition.

COMPLICATIONS OF RESPIRATORY INFECTION

Common respiratory illnesses can cause complications in persons with ALS. To reduce your odds of infection, try to avoid contact with people who have colds or flu. Be sure to practise regular hand-washing.

Bronchitis

Bronchitis is an inflammation and irritation of the **bronchial tubes** in the lungs characterized by a cough. This condition may be caused by viruses, bacteria, environmental irritants such as cigarette smoke, or allergies. In response to the inflammation, airways secrete a sticky mucous. If a person is unable to produce an effective cough to clear the lungs, there is a risk for developing pneumonia.

If you do get a fever with thick discoloured, or bloody mucus coughed from the chest, or if you notice that the mucus has changed from clear white to yellowish,

tell your doctor immediately. These could be symptoms of a respiratory infection that could lead to pneumonia.

Pneumonia

Pneumonia can be caused by bacteria or viruses, and may be a complication of a cold, flu, bronchitis, or aspiration. Pneumonia causes the very small air sacs in the lungs (alveoli) and surrounding tissues to fill with inflammatory cells (pus) and mucous which prevents the normal exchange of O₂ and CO₂. When this occurs oxygen is not taken adequately through the alveoli into the blood and a low oxygen level may result.

Aspiration

Aspiration occurs when liquids or solids get into the airways and block airflow and gas exchanges (O₂/CO₂), cause irritation, or carry infection from the mouth and gums. Small amounts of aspirated material can be coughed out. If aspiration is frequent and involves large amounts of material, an evaluation by a swallowing specialist (speech language pathologist with expertise) and respiratory consultant is advised. Changes in nutrition and swallowing techniques may be recommended.

STAYING HEALTHY: PREVENTIVE AIRWAY MANAGEMENT STRATEGIES

Preventative airway management measures initiated at an early stage can prevent unnecessary hospital emergency visits and intensive care unit admissions. Airway management strategies are used to either enhance your ability to recruit (take in) air volume into the lungs or cough effectively. Strategies range from self-care techniques to the use of assistive devices. Incorporating airway management strategies can promote better lung function longer, and thus promote quality of life.

Lung Volume Recruitment (LVR) and Assisted Coughing Strategies

Before trying any assistive breathing and coughing techniques, make sure to discuss them with your healthcare professional. Some exercises and techniques are more complex than others and require training and practice.

For more information about many of the techniques, talk to a **respiratory therapist, respirologist, or physiotherapist**, as well as visit The Rehabilitation Centre web site <http://www.rehab.on.ca/program/respiratory/neuromuscular.html>. Baseline pulmonary function and a respiratory assessment are recommended.

- **Deep Breathing Exercise**

Exercises to use full lung capacity can compensate for weakened muscles. This can be achieved with the following deep breathing exercise:

1. Sit at a table, hunched over slightly with your weight supported on your hands or elbows. This position expands the rib cage for larger breaths by allowing your shoulder and neck muscles to assist your breathing.
2. Breathe in as deeply as possible, hold the breath for a few seconds, and then exhale.
3. Repeat several times.

Some doctors believe that patients who do deep-breathing exercises are less prone to lung infections and partial lung collapse. Ask your physiotherapist to demonstrate an exercise routine and advise you about the use of exercise devices.

- **Assisted Cough Techniques Without Devices**

Assisted coughing is a technique where a caregiver applies forceful pressure to the abdomen and occasionally the chest wall, timed to the effort of coughing. This action assists the weakened muscles responsible for an effective cough. **These techniques are very helpful, effective and easily learned. They are very empowering and relieving for caregivers who, without them, feel powerless to assist their loved ones in their respiratory distress.**

1. **Abdominal thrust:** Just immediately prior to a timed cough effort the caregiver applies rapid and forceful pressure to the upper abdomen in an inward and upward fashion between the bottom of the breast bone and the navel (umbilicus).

2. **Lateral costal compression:** Sometimes combined with the abdominal thrust, or used alone when a PEG tube has recently been introduced. Firm but not too forceful pressure is applied by each hand on the lower rib cage below the breasts or at the sides of the chest timed to a cough effort.

3. **Self-assist:** This is most appropriate for paraplegic patients and would rarely be used by ALS patients. The individual in a stable wheel chair takes a full breath in, folds their arms across their abdomen and forcefully pitches their torso forward over their crossed arms. Once a significant pressure has been generated in the abdomen and chest they release the glottis and cough.

- **Assistive Cough Techniques With Devices**

1. **Modified resuscitation bag:** A special hand-held bag is usually used for resuscitation (start up breathing again) in a respiratory or cardiac arrest. This useful tool can be modified with the addition of one-way valve tubing and a mouth-piece (or mask) to allow the passive introduction (by one's self or a caregiver) of volumes of air into the lungs. This allows re-achieving volumes which have been lost due to muscle weakness. Its effectiveness is directly dependent on bulbar function. Once a larger volume has been accommodated then an abdominal thrust (as above) can be added to enhance the effectiveness of the cough.

2. **"CoughAssist" (Mechanical in-exsufflation machine):** This is a very effective device for those who maintain sufficient bulbar function but have a weak cough. It is a vacuum that allows attachment through a face mask to both the positive pressure side (to help inflate the lungs) and the negative pressure side of the vacuum (to fill up the lungs and then forcefully empty them to mimic an effective cough). The high speed expiratory flow is effective in clearing airway secretions. Considerable experience is required in the assessment and introduction of this device and it is quite costly.

3. **Volume ventilator:** For people who are already using a volume ventilator for mouth-piece ventilation, the volume of an assisted breath can be held by the glottis and added to the next machine-delivered breath achieving a breath-stacking effect.

This improves both the volume (range of motion) of the lungs (and thorax) and the effectiveness of the cough. Again this is most effective with maintained bulbar function.

Lifestyle Strategies

- **Do not smoke**

In addition to all of the other problems smoking causes, it reduces lung capacity. It can also cause increased phlegm that some patients will have difficulty clearing from their airways.

- **Be cautious with alcohol and sedatives**

Alcohol and sedatives may increase the risk of aspiration or hypoventilation during sleep.

- **Avoid allergens**

If you are allergic to pet dander or dust mites, make sure your home is free of pets that cause you problems and is regularly dusted. Investing in an air purifier with a HEPA filter can help keep the air free of particles that irritate your airways.

RESPIRATORY FAILURE: ADVANCED BREATHING MANAGEMENT OPTIONS

Failure of the respiratory muscles is an eventual result of ALS. The breathing muscles become too weak to expand the lungs and to provide an adequate cough, thus requiring assisted breathing support. The decision of whether or not to use breathing support is yours, but it is a decision you should make only after consulting with your family, doctor, and other healthcare team members. The choice to use breathing support will have important consequences for your way of life and that of your family and caregivers. You will have to decide whether you are willing to accommodate your life and that of your family to the potential dependence on a breathing support apparatus. **This decision must be made well before life-support ventilation is needed.** It is also important to have a clear understanding of what is covered by your available health insurance/benefits. **Breathing support can be provided non-invasively or invasively.**

Mechanical Ventilation: Things to Consider

Type of Ventilation	Non-Invasive Ventilation	Invasive Ventilation (with Tracheostomy)
Advantages	<ul style="list-style-type: none"> • Nose and mouth offer convenient routes for the delivery of breathing support and therefore do not involve any kind of surgical procedure • Easier to use than invasive ventilation • Usually more comfortable • Associated care is less complex and with fewer complications • If using a mouth piece with the volume ventilator your cough will be more effective • Longevity may be increased • Less costly overall • Little or no problem with aspiration • With appropriate lung hygiene regimen, lung infection occurs less frequently • Easiest to stop therapy 	<ul style="list-style-type: none"> • More secure system if you are ventilator dependent • Has been found to provide much longer survival • No interface required therefore the face free of headgear, straps, and skin pressure problems • Doctors, nurses, and respiratory therapists tend to be more familiar with tracheostomy care and invasive ventilation
Challenges	<ul style="list-style-type: none"> • Initially, finding the optimal interface selection may be challenging, specially with upper extremity paralysis • Custom-fitted silicone-molded nasal masks which may provide a better seal at higher pressures, require additional time and expertise • Some people with ALS find it difficult to adapt • If bulbar impairment is severe, non invasive breathing support may be more of a challenge to use • Gastric distention may occur • Most centres are familiar with traditional invasive ventilation however, few have experience with day time mouth piece ventilation 	<ul style="list-style-type: none"> • Some people feel it is too invasive, and increases their disability and dependence • The tracheostomy tube is a foreign object in the body thereby increases secretion production and infection occurrences • Secretions require suctioning with a catheter through the tracheostomy, during the day and at night which is uncomfortable • Coughing to clear the airways is difficult • The tracheostomy site (stoma) can become infected, bleed, or develop granulations that need to be removed • Associated care is more complex than non-invasive ventilation, therefore requiring more skills • Some people have difficulty with speech and swallowing • More costly overall

Non-Invasive Ventilation

Non-invasive ventilation is achieved with a bi-level unit, a positive pressure volume ventilator, or much less commonly, a negative pressure ventilator. **The most common breathing support is the bi-level (commonly known brand name product is the BiPap™).** The bi-level helps you breathe by providing two levels of air pressure. A higher pressure when you inhale assists weak inspiratory muscles and a lower pressure when you exhale. A circuit of tubing is used to deliver air from the bi-level to your interface (a mask over nose or nose and mouth).

Non-invasive ventilation is often initiated at night only. The practice of daily non-invasive lung hygiene techniques such as lung volume recruitment with assisted cough is a must when opting for non-invasive ventilation. As the disease progresses increased breathing support will be required. This may be provided by using a mouth piece during the day with a volume ventilator and a different interface for night time use. Your respiratory therapist will assist you in finding the appropriate interfaces.

Although some individuals using non-invasive ventilation may eventually rely on it 24 hours per day this is very difficult and cannot be sustained for extended periods of time. When someone does require non-invasive ventilation greater than 16 hours per day, it is likely they will soon need to make a decision about whether or not they want to continue with mechanical ventilation requiring a tracheostomy.

Invasive Ventilation

Invasive ventilation requires a **tracheostomy**, a surgical procedure. The surgeon creates an opening in the neck connecting directly to the windpipe or trachea. In the surgical opening a curved plastic tube is put in place. This tube is connected to a hose and volume ventilator, which assists with breathing. Whenever necessary, a suctioning catheter can be inserted into the tube in the trachea to remove secretions. Most patients with a tracheostomy have a feeding tube (PEG tube) as well.

Prior to making a decision about invasive ventila-

tion both you and your family should learn from a doctor about the full implications of invasive ventilation. This will require full time, 24 hour support from trained caregivers. Can you count upon the assistance of family members? It is a full-time job for three people, assuming eight-hour shifts. Provided you have limited bulbar involvement speaking can continue through the use of a tracheostomy speaking valve such as the Passy-Muir® valve

Making Your Decision About Ventilation

In summary, it is best to learn what you can about ventilation options early on in the disease prior to an emergency. Planning ahead enables you to be in more control. **Communicate your wishes regarding use of a ventilator in a personal healthcare directive (Durable Power of Attorney)** and ensure that your advocate is agreeable to supporting your decisions. Be sure to provide your doctor with a copy of the directive and inform your family members where you keep it at home. Without a directive and making your wishes known, you can probably assume invasive ventilation with tracheostomy will be initiated in the event of a respiratory crisis. If at any time you decide you are interested in any form of breathing support, you should be referred to a respirologist.

Final Note on Breathing Management...

If you didn't know before reading this section, you now know breathing management plays a major role in ALS management and enhancing quality of life. Monitoring your breathing function is very important for identifying problems early. Early detection is critical because there are things you can do to reduce risk of infection, promote better ventilation for longer, and possibly extend your survival. Many decisions about interventions may need to be made along the way. Having good information about what is available to you and discussing options with your family and healthcare professionals will help you to make decisions that will be best for you.

Weakened mouth and throat muscles, weakened hands and arms to brush teeth and tube feeding are all changes which require special treatment for mouth care. Food can easily collect in pockets in the mouth as well as between teeth. Retained food debris, thick mucous and decreased saliva all contribute to poor oral conditions: too much plaque and tarter, halitosis (bad breath), gingivitis and tooth decay. It is important that you take care of your mouth to prevent harmful bacteria from developing. Bacteria can cause bleeding gums, bone loss, mouth and tooth infections.

There are many products available to make oral care easier. Assistance with oral hygiene can greatly impact one's self esteem and dignity.

ORAL HEALTH ISSUES

Here are some issues that you need to be aware of and tips on promoting good oral health.

Going to the Dentist

Not many dentists will know about ALS, its effect on your ability to keep your teeth clean, excess saliva or the precautions they should take because of a patient's swallowing problems. Make your dentist fully aware of these problems. Continue with regular dental cleanings for as long as possible at your family dental care office. Consult your dental hygienist for preventative home care instructions.

Mouth Freshness

An applicator with lemon and water can be used to keep the mouth fresh. You can also try an oral rinse such as Biotene or non-alcohol mouthwashes.

Furry Tongue

Some individuals with ALS with bulbar symptoms develop a furry tongue. Water may be used to clean the tongue with an applicator or washcloth. If the tongue appears white for any length of time, consult your doctor or dentist. Sometimes a condition called 'thrush' can develop. It is a fungal infection and is easily treated.

Saliva Build-Up

Build-up of saliva is a common problem among people with ALS who have tongue and throat muscles that are weak and not able to automatically swallow the saliva that builds up in the mouth. Very thick mucus can also build up in the mouth, making the problem worse. This build-up of saliva can cause choking and disrupt sleep.

Relief may come from home remedies, over-the-counter products, and prescription drugs and, in extreme cases, even surgical procedures. Advice should be obtained from your doctor and/or your local pharmacist. A side effect of drugs to relieve a build-up of saliva is "dry mouth." It's a question of finding the right balance for you.

The following have proven helpful for managing saliva:

- Portable suction machine (check with your doctor before purchase as suctioning may be counter-productive in ALS)
- Decongestants and over-the-counter medications used for colds and allergies
- Antidepressants - Amitriptyline may reduce saliva and enable sleep
- Anticholinergic drugs to reduce spasms of smooth muscle in the bronchi and decrease gastric, bronchial and salivary secretions
- Transderm V patches usually used for motion sickness also tend to dry up saliva

The following have been shown to be helpful for treating thick mucus:

- Apple juice, grape juice, or hot tea with lemon
- SSK1 (potassium iodide) 10 drops in a glass of water two to three times per day. This may take one to two weeks to be effective. A prescription is required
- An expectorant cough syrup
- Meat tenderizer mixed with a little water can be used to coat the tongue or can be placed under the tongue
- Papaya extract placed on the tongue may prove helpful

Precautions: Some of the suggestions for saliva management include use of medication. Here are some important points to keep in mind when taking over-the-counter medication:

- Always first talk to your doctor or nurse about medication recommendations before use
- Sleepiness is often the side effect of medications
- Beware that anything containing alcohol can increase muscle weakness, at least temporarily
- Beware of anything that slows breathing
- Before putting any medication, crushed or otherwise in a feeding tube, check with a pharmacist to ensure that it won't harden and clog the tube
- Generic products are usually cheaper

Excess saliva has one advantage! If you have excess saliva and are still able to eat by mouth, mixing more saliva with your food makes it both easier to swallow and digest.

Dry Mouth

Although management of saliva is common in many people with ALS, especially those with difficulty swallowing, some are troubled by excessive mouth dryness. A dry mouth can cause thick mucus to form, which may in turn cause serious choking problems.

Excessive dryness is usually caused by one or more of the following:

- Breathing mostly through the mouth, rather than the nose can dry out the lining in your mouth. If you consistently wake up with a dry mouth, thick mucus and/or a sore throat, you may be breathing through your mouth while sleeping
- A side effect from some surgical operations
- Stress from chronic anxiety or depression
- A side effect from some medications
- The natural process of aging
- Smoking

The following have proven helpful for a dry mouth:

- Make a conscious effort to breathe through your nose
- If you suffer from nasal congestion, ask your doctor for assistance (nasal congestion leading to a dry mouth can cause thick mucus to form, which in turn may cause choking problems)
- Increase your liquid intake (ask your speech pathologist and dietitian which consistency is best for you)
- Use a vaporizer (clean the vaporizer once a week with vinegar and water)

- Ask your pharmacist about oral rinses, artificial saliva spray, and tablets

ORAL HEALTH DEVICES

Toothbrushes and Flossers

If you have weakened hands and arms, an **electric toothbrush** is ideal for brushing your teeth. There are a variety of electric brush shapes and price ranges: Oral B, Philips Sonicare, Colgate and Crest spin brushes. Others may find that a toothbrush is very helpful for removing build-up, which occurs on the teeth, particularly after tube feeding. There are many brushes widely available and a soft bristle brush is always recommended to minimize tissue trauma. Newer to the market are both **manual and electric flossing aids**: Reach Access, Butler Floss Handle and the Oral B Humming Bird Flosser.

- **Proper Brushing Technique:**

1. Place the toothbrush at a 45-degree angle to the gum-line.
2. Use a gentle circular motion several times in the same spot.
3. Finish with a sweep of the brush from the gum-line to the chewing surface of the tooth.
4. Repeat this method as you brush from the right to the left side of the mouth.

Also be sure to brush the inner surfaces of the teeth. If a caregiver is brushing for you, the handle of a soft grip toothbrush could be used as a mouth prop to assist access.

Suction Unit

Another method for moving food caught in pockets of the mouth is a suction unit, which is also used for removing excess saliva to reduce drool.

Final Note on Oral Health...

As noted earlier, many dental care professionals may not be familiar with ALS and symptoms of the disease that will impact dental treatment.

You are encouraged to bring this section of the manual with you to your dentist or hygienist. Educating them will improve their ability to work with you to promote optimal oral health and treat problems if they occur.

Helpful Web Sites:

www.cdha.ca

www.colgate.com

www.crest.com

www.dentalresoursenet.ca

www.prodhhelp.com/ca/oral.shtml

www.jbutler.com

www.oralb.com

www.oral-care.com

www.jnj.com/hom.html

Conditions, such as ALS, often described as fatal, or terminal, could perhaps better be described as "life-shortening." The only true fatal condition there is, is life itself. End-of-life issues such as the kind of care you want and organizing financial and legal affairs are things all of us need to address no matter what the status of our health is today. However, unless we are knowingly in our last phase of life, most of us would prefer not to talk about it, much less plan for it.

How we deal with death can be deeply affected by our cultural and religious values. Some cultures view death as a welcomed spiritual transition. Others believe that death and rebirth are a continual process. During our lives, the cells in our bodies die at an ever-increasing rate, while at the same time our learning, intelligence and spiritual awareness can continue to grow.

For many, the final stage of life can be a time of tremendous personal growth previously not experienced at any other time. Accepting that time is limited can motivate one to really make a difference in one's life and those around them. This time of life can be a very special gift. Approaching the end-of-life phase in a positive, well-thought out way, can make for a good death which can help provide comfort to surviving friends and family members.

The information in this booklet encompasses end-of-life concepts, advance care planning, and ideas for telling one's life story

WHY IT'S IMPORTANT TO TALK ABOUT IT

As ALS progresses, there are numerous practical issues to deal with and arrangements to be made. It is often difficult for a family to talk openly about these practical matters. At some point someone has to deal with financial changes, tax issues, insurance, medical and personal care choices, a will and funeral arrangements, as well as distribution of assets and belongings. It is best to discuss and plan for the decisions you need to make well before death is near. **Poor communication and planning of these practical concerns could force family members to make decisions without knowing your wishes,** which could undermine relationships, cause unnecessary financial hardship, or result in a less peaceful, comfortable death.

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APPROACHING THE CONCEPT OF END-OF-LIFE

What Does End-of-Life (EOL) Mean?

In healthcare terms, end-of-life is the stage in a person's life where death is expected within a short period of time (e.g., within six months or less) barring the use of artificial life support interventions.

In personal terms, end-of-life may begin at diagnosis for some and not until the final days for others.

Regardless of when you believe the end-of-life stage is, making decisions about the kind of care you want at the end-of-life and the provisions you want to make for your surviving family members should take place much earlier.

Contemplating the End-of-Life Phase

- **Acceptance**

Accepting that life is coming to a physical end may be less difficult when the illness has been lived with for some time. With ALS and the functional and role changes associated with it, lifestyles often slow down in pace, perhaps giving one more time to reflect. When diagnosed with ALS, a future fraught with changes and losses is faced, and thus an ongoing grief process is experienced. Acceptance is one of the stages of grief one passes through along the journey. Sometimes we accept things, then go back to denial, or anger, and eventually, work our way back to acceptance. Because for many people with ALS there is time for this process to unfold, there is time to contemplate what lies ahead.

Instead of looking at oneself as dying, it may be easier to accept being in the final stage of life.

Therefore, the concentration is on living. Once one accepts that the natural progression of their illness is a shortened life, their acceptance can help prepare and comfort those around them. A sense of peace about death can open up opportunities for deep and meaningful communication and planning for what is to come.

- **Expectations of the Dying Process**

An area that is most frightening to people who are facing death, and those around them, is what actually hap-

pens during the last days and hours of the dying process. To minimize fear, it is best to **ask your doctor and nurse what to expect of the dying process and if there are specific expectations for someone with ALS** as opposed to someone without. If you have contact with a palliative care/hospice doctor or nurse, or pastoral care professional ask them to describe to you what the dying process will be like and what supports will be there for you and your loved ones. Have the family members you expect to be with you be part of the discussion. They may have fears and questions they will also want to address. A video that may also be of help, "**Facing the Fears-Making the Journey**," is available through the Canadian Hospice and Palliative Care Association (<http://www.chpca.net>).

Also ask your doctor or nurse what factors may impact your comfort during the dying process. The answers you get to your questions will help you make decisions about specific aspects of care you want documented in your advance directive to ensure a "good" death.

- **Experiencing A Good Death**

"He died peacefully." Undoubtedly, we have all heard this phrase. Unfortunately, we have also heard of other stories where people did not die quite so well. With gaining momentum and acceptance of the palliative care and hospice movement in our society, we hope to hear more stories about "dying well." The goal of palliative care is providing comfort. Dying well also includes experiencing a death that is dignified by fulfilling expressed health and personal care wishes of the dying person.

We can take control in advance to promote a good death for ourselves. Some of the things that can help are:

- Making peace with family and friends if needed
- Exploring spirituality and faith
- Completing a living will and advance directive and sharing it with family, doctor, and clergy
- Communicating openly with loved ones about the desired environment for the last days or hours of life

ADVANCE PLANNING

Advance planning can avert potentially difficult situations for family members. Limiting the number and type of decisions others have to make on one's behalf can substantially reduce their stress and tension levels. However, once plans have been made, decisions should be revisited every so often to make sure the plan is still desirable and to make changes if necessary.

Advance planning can be started as soon as you are ready, and may include choices no different from before you had ALS. Regularly reviewing your advance directives to reflect your current outlook allows you, your proxy decision maker, and the health-care team to become comfortable with your choices.

Legal and financial planning information is discussed in detail in **Section 6, *Legal and Financial Considerations***. The two advance planning areas of focus in this sub-section are: **end-of-life care choices** and **bereavement planning**.

End-of-Life Care Choices

Explicit instructions in a **living will (advance health-care directive)** should help to ensure a dying person is treated according to their wishes and will have a dignified death as defined by them. Completing a living will and appointing a substitute decision maker ("proxy) in a legal document, frequently called a "**power of attorney for health care**," in the event one is unable, or too sick to communicate their wishes for care is critical. Wishes expressed in a living will can be as specific as the individual wants them to be. When preparing how you want to be cared for in your final phase of life **consider not only what medical care measures you want, but also where you want to die and who and what you want to have around you.** This is your life *and* death, and you are entitled to being in control of your dignity. More detail and references for advance directive forms are included in **Section 6 *Legal and Financial Considerations***.

In a nutshell, there are four steps to advance care planning:

1. **Gather** information.
2. **Talk** about decisions.

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3. **Prepare** and sign an advance directive.
4. **Inform** appropriate others and provide them copies.

- **Medical Care Choices**

The type of medical care that you want during the course of your disease as well as in your final days is up to you and no one else. Just as it is you who needs to make the decision about a PEG tube or a ventilator at given stages of your ALS progression, it is you who will decide when to suspend these treatments.

Decisions about medical interventions geared to extend life are very personal and should be based on medical information, financial resources, caregiver support and resources, and your perceived quality of life.

It is critical to periodically review advance care plans. While quality of life means something different to every single person, it can also change meaning within an individual over time. For instance, a person with ALS may indicate at one point that in the event of respiratory failure they do not wish to use mechanical ventilation. However, that same person may change their mind when they hear there is a new grandchild on the way and that mechanical ventilation in the event of respiratory failure would ensure being able to live and greet the new member of the family. On the contrary,

someone with a PEG and/or ventilatory support may decide they no longer want to live with those interventions if a given situation were to arise. **It is important for professional caregivers to periodically review their patients' advance directives with them** and discuss what they are presently feeling about their choices.

Three major intervention issues that ought to be addressed in a directive are:

Resuscitation: Ask yourself if you do or do not want to be resuscitated in the event of respiratory failure. Ask your doctor about how you go about getting a Do Not Resuscitate (DNR) order, if that is what you prefer.

Life-extending interventions such as feeding tubes and mechanical ventilation: A sample advance directive that focuses on PEG and mechanical ventilation, designed by the University of Washington for patients with ALS, is referenced in **Section 6, *Legal and Financial Considerations***. This document may help you make your decisions and assist you with wording to use if you are required to use a specific form recognized by your Province.

Pain management: Stating one's desire to have pain appropriately monitored and treated is very important. This should be the goal of care, but it is documented that many people die in pain unnecessarily, especially when they are not under the care of palliative or hospice care specialists.

In discussing the content of the advance directive with your healthcare team, it is vital you discuss the medical management and strategy of care of symptoms when the conventional intervention is not desired (e.g. invasive ventilation). In particular, when you choose not to have breathing support, you need to have explicit strategies for managing feelings of breathlessness, and for the rare instance of acute breathing distress. Faced with the dilemma of acute breathing distress and yet a directive indicating no tracheostomy tube, it may be reasonable to accept temporary breathing support to control symptoms. Artificial ventilation can be withdrawn at any time, and the medical team can ensure that this is done comfortably. It is legally and morally acceptable in Canada to ask to have

When you prepare a healthcare directive remember to:

1. Discuss your decisions with your loved ones and doctor.
2. Keep a copy in a safe, easily accessible place at home that your caregivers know about.
3. Give a copy to your doctor for your medical chart and your appointed decision-maker(s).
4. Carry a wallet sized card with you that names your appointed proxy and their contact information, in the event of a medical emergency outside the home.
5. Revisit your decisions every few months and re-evaluate. Communicate any changes you want to make.

life-sustaining therapies including mechanical ventilation withdrawn. However, this should be discussed specifically with your spiritual advisor, as there are some religions where withdrawal of mechanical ventilation is not acceptable.

- **Choices About Where to Die**

Many people with ALS prefer to die in the peace of their own home. Others may prefer to be in a facility of some kind. Whichever is your wish, you must communicate this to your family while you are still able to do so. Factors that might not make dying at home possible include your family's financial or emotional inability to cope, caregiver burnout, respiratory problems or insufficient home care services. Remember, calling 911 will bring a medical team obligated to keep you alive, including artificial life support that you may not want. Some provinces may have a death at home policy which allows you to by-pass calling 911. There will be forms to complete. Please check with your local ALS Society.

If you choose to die at home, you may consider some specifics such as:

- Which room do you want to be in?
- Is there a favourite view you want to be positioned toward?

- **Choices About Who and What You Want Around You**

To take the guess work out of deciding who is appropriate to visit with you during your last days or hours, tell your caregivers ahead of time.

A few of the questions you need to ask yourself and answer include:

- Are there people you want to re-connect with now?
- Whom do you want near you in your final days?
- Is there something special you want them to do for you, like read your favourite book aloud?
- Do you want your pet at your side?

Some people are very specific about how they want the environment around them to be. It is a good idea for you to contemplate the kind of setting you think would put you at ease. Communicating this

to caregivers and other loved ones will enable them to make sure you have the setting you want. It will also make them feel good about being able to help you feel at peace and "do something" constructive.

Some of the questions you may ask yourself include:

- Do you want music playing and if so what kind?
- Do you want to be near an open window so you can feel a breeze?
- Would you like candles with a favourite scent burning?
- Are there important pictures of places you've been, or family gatherings that you want in your view?

There are many decisions you can choose to make.

Don't feel overwhelmed. Give as much thought as you can and involve others to help you sort through it all. **There are some excellent resource books and videos available to support you.** Many are listed in the "Resource Section" of the *Manual*. One in particular is *Living with Life-Threatening Illness: A Guide for Patients, Their Families, and Caregivers*, written by *Kenneth Doka, PhD*, a leading authority on end-of-life issues. Check to see what may be available through your local ALS Society. What they do not have they may be able to help you find. An excellent and very comprehensive resource guide which includes numerous references to web sites, books, and videos can be found on-line at www.lastacts.org.

Bereavement Planning

Another type of advance planning is bereavement planning. It is very beneficial for family members and close friends of a dying person to **identify resources that will help them cope with the overriding sense of loss and grief they will experience after the death.** One such resource is the booklet, *Coping With Grief*, published by the ALS Society of Canada which is available through the ALS Society in your province. This booklet also touches on **anticipatory grief** which is experienced before death by both the person who is going to die and their loved ones. This type of grief is of course unique to those affected by long-term illnesses. During the course of the illness there is grieving around losses of function, roles, and hope. Triggers of anticipatory grief could be a terminal diagnosis, decreasing control over daily activities, loss of function, changes in roles, and lost hopes and dreams

for the future.

Psychosocial support during the grieving process can help individuals process and express their feelings in healthy ways that will eventually help bring about healing. Sources of support in the community include **hospice organizations, members of the clergy, and mental health practitioners** such as therapists who specialize in grief counselling. Another great source is **peer support** from others who have gone through the same experience. Your local ALS Society may be able to match bereaved caregivers with former ALS caregivers who either meet as a group or are willing to help others one-on-one.

LEAVING A LEGACY

Have you ever thought about how you want to be remembered, or what you want to leave behind to help comfort your loved ones? **Telling one's life story and documenting memories on paper, video, or audio tape can be an excellent way to help transition to the final stage of life and leave one's mark on the world.** The process can be very therapeutic in terms of searching for the meaning of one's life and identifying core values and beliefs. Sharing it will pass on valuable lessons to others.

Looking back and sharing special memories can be both a fun and enriching experience for yourself as well as loved ones engaged in the process. Family and friends will likely be able to trigger memories of things you may have forgotten, or buried and vice versa. Leaving a legacy may spark feelings of satisfaction of a life well-lived as well as give survivors a tangible memory of who you are, what you believed, and how you lived your life to inspire future generations.

If you choose to leave a legacy of memories, there are many formats you can use. If you can type or use computerized communication software, documenting a written story may be your preference. If your speech is good, you may prefer to have a friend or family member document or videotape your stories as you tell them. It's up to you. An excellent resource for getting started and guiding you through the process is, *A Guide to Recalling and Telling Your Life Story*, published and available through the Hospice Foundation of America (<http://www.hfa.org>). This is a work book that suggests topics such as "Family Life," "Growing Older," and "Reflections"-and suggested

questions to use to elicit stories and experiences from you and your loved ones.

While you may need to spend a few dollars to buy the *Guide* plus some documentation materials, what you end up with is a rich personal legacy which is priceless.

Final Note on End-of-Life Issues and Advance Care Planning...

Everyone recognizes that end-of-life issues are difficult to face and talk about. However, keep in mind that avoiding advance care planning can be more difficult later for your loved ones if they are left making decisions for you. We have all heard stories about families that were ripped apart over differences of opinion about what they perceived the dying person would want. Try to approach the end-of-life phase as an opportunity for reflection, planning, and enrichment.

Assistive equipment plays a major role in the lives of persons with ALS. Because ALS is a progressive condition, physical changes occur over time requiring the on-going need for assistive devices to maintain functionality. ALS will affect each individual somewhat differently. For you, a wheelchair may not be necessary. For someone else, specialized communication devices may never be required. **This section attempts to give you an overview of the many types of equipment that may be needed by someone with ALS. Not everyone will need all of it and certainly not all at once.**

BEFORE PURCHASING EQUIPMENT

Before purchasing assistive equipment or modifying your home, you are advised to **consult with professionals who are familiar with the advantages and disadvantages of the equipment that is available to serve your needs.** Equipment can be expensive, so you need to make wise choices.

Think through what your needs are, both in the immediate and longer term. Discuss your needs with a doctor, nurse, physiotherapist, occupational therapist or other healthcare professional with knowledge of your condition, and familiarity with the available equipment to serve those needs. **Make sure to check your own private or group insurance plan to determine what coverage you have** and what documentation you require for coverage. Some plans may only cover certain items every so many years.

Find out from healthcare professionals, in particular social workers what loan equipment or financial assistance might be available. Your provincial ALS Society is also an excellent source for helping you locate the kind of equipment you may need. In many cases, the Society will have the equipment you need through their equipment loan program.

You may also want to visit local suppliers to test out various equipment options.

- **Check Into Financial Support Programs**

Check with your local healthcare professional and equipment suppliers to find out what government support programs are available in your area for equipment, and for home renovations required to accommodate your disability. Some programs will fund only one project per patient. Therefore, consider carefully what your longer term needs are likely to be before applying for this type of assistance program.

- **Check with the ALS Society for Equipment**

Some ALS Units and Chapters in your area may have a limited or extensive inventory of equipment acquired through the generosity of contributors and for use by people with ALS. See the information provided by your provincial ALS Society for more specifics.

- **Identify Local Equipment Suppliers**

Most suppliers of equipment for people with disabilities are listed in the phone book under "Hospital Equipment and Supplies."

- **Get Advice from Home Care**

Home care staff is another source of information about assistive equipment. Many of them have probably visited other people with similar needs, and have seen how useful different types of equipment have been.

TYPES OF EQUIPMENT

Assistive Tools for Activities of Daily Living (ADL)

Many people with ALS develop trouble with grasping and manipulating objects. It is often possible to **modify everyday tools** or to substitute specially designed versions of such tools to compensate for weakness in the muscles of the fingers, hands and wrists.

For example, **a knife, fork and spoon with extra-thick handles can make eating much easier.** There are also sets of cutlery available with thick, long handles that compensate to some extent for impaired shoulder movement. **Mugs with oversized handles** will allow you to slip all fingers under the handle,

reducing the danger of spills. A **plate guard** gives a slight vertical edge to a plate so that food can be pushed against it onto a spoon or fork.

The double action of gripping and turning a doorknob may also be a problem for you. A **doorknob adapter** could be the answer since it allows the door to be opened by pushing down or pulling up the lever. A fat wooden or plastic handle attached to your door key can help with the turning motion necessary to turn the key in the lock.

To assist with written communication, use **thick pens or pencils** that are easier to grasp than the usual thin variety, or use a writing aid that consists of a block holding a pen or pencil.

Clothing fasteners can be difficult to use if your hands and fingers are weak. There are devices to assist in fastening buttons. **Velcro** is a popular replacement for both buttons and zippers on clothing. Shoe closures such as buckles or laces can also be replaced with Velcro closures by a pedorthist or shoemaker/repair shop. Best of all, are such items as pants or skirts with **elastic waistbands** need no fasteners at all. Although pullover tops have no closures, they can be some of the hardest clothes to get on and off.

Electronic switches, known as "**environmental controls**," can be installed in the home to enable a person with ALS to turn control lights, radios, televisions, open and close doors, start a coffee maker, etc. with the palm of the hand, a head movement, or even a puff of breath.

Telephones that do not require the use of hands are also available.

We have mentioned just a few of the modified or special tools available to make everyday life easier for you. There are many more such tools that your occupational therapist can tell you about. It is important to discuss any tool with your therapist before making a purchase. Some devices are reasonably priced and some are expensive. Some will assist you for a long period of time, whereas others may only be use-

ful in the short-term. Your therapist can save you disappointments as well as costly errors.

Body Supports

The various types of body supports available are called **orthoses**. Available through hospitals and clinics, orthoses help to support your joints in certain positions when your muscles weaken, prevent contractures if spasticity is a problem, enhance comfort, and aid in function. Orthoses are typically prescribed by a **physiatrist**, and may be recommended by your doctor or physiotherapist.

There are more orthoses available than we can discuss in this brief overview, but some of the major ones are described.

- **Foot and Ankle Supports**

Many people with ALS experience "foot drop" which is caused by weakened muscles supporting the ankle joint. This leads to stumbling on stairs or curbs and tripping while walking. The answer may be a simple ankle-foot splint made of plastic that is inconspicuous under trousers or slacks. These devices are often referred to as "**ankle and foot orthoses**" (AFOs). For more information on AFOs, talk to your doctor.

“ Using short stemware, preferably the heavier glass type can be easier than holding a cup or glass. ”

Terry Schwartz
ALS family caregiver

- **Hand and Wrist Supports**

A wrist and thumb splint can stabilize the wrist and thumb, helping you to grasp eating utensils and other objects. Additional supports can be added for positioning the fingers, enabling you to make finer movements with weakened fingers, such as are required for writing.

The **universal cuff** may be the most familiar orthotic aid. This straps over the hand and allows you to grasp such objects as cutlery, hairbrushes and other small personal objects. A thumb splint helps in squeezing the fingers in opposition to the fingers and makes it easier to grasp and hold an object for use.

- **Shoulder and Neck Supports**

Weakened shoulder muscles can cause the arms to

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"hang" more than normal, which can become very painful. A shoulder sling can pull the arm up, reducing the pressure on arm muscles and ligaments. There are also cervical collars that can support the head when neck muscles are weak.

Walking Equipment Aids

- **Canes and Walkers**

Most people with ALS will require a cane or walker at some point. These are especially useful when one leg is stronger than the other. Canes should always be used on the stronger side, with the cane moving forward with the weaker leg. There are single or multi-legged canes, so discuss with your physiotherapist which type of aid might work best for you. Another type of cane extends up the lower arm, almost to the elbow, with a ring that fits around the arm for added support. Walkers provide maximum support because they spread the weight over a wide area. **The choice of cane or walker should be made in consultation with your doctor and physiotherapist and you should not try to use it until you have received instruction from your therapist.**

Wheelchairs

The decision about when to get a wheelchair is one that you will make with your doctor. A wheelchair will be prescribed for you by your occupational therapist.

There are different types of wheelchairs and wheelchair features that require consideration. Straps may be needed to hold in arms and feet, and a seat belt is also a good idea. Hand-rim pegs are helpful if you have trouble gripping with your fingers, but have strength in your arms. There are wheelchairs that have a double hand rim for operating from one side which is useful if you have strength on one side only. Anti-tipping bars may be extended out the front or back to prevent the wheelchair from tipping forward or backward. A commode attachment is available when you are alone for long periods. Tray or table-like platforms should be a standard feature that can be attached to your wheelchair. Folding wheelchairs are useful because they can be put in a car. Ask the professionals who help you with your wheelchair decision about other available features.

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For the more advanced stages of ALS, a battery-powered wheelchair may be required. These can be operated using a joystick or other switching devices that can be controlled by almost any part of the body that you can move. Control switches can be modified to operate with even very small muscle movements.

Getting fitted with a wheelchair requires careful consideration and professional advice. First, your size will determine the appropriate height, depth and width of seat, as well as the height and width of the back. An occupational therapist can help you get the right chair for you.

It is very important that you get the right chair.

Many factors will have to be considered, such as your physical condition now and in the future, your financial situation, insurance coverage, the availability of financial assistance, your level of help from family and friends, and the sorts of services your community provides for transportation. A wheelchair is a major purchase, and if it is going to enhance your life now and in the future, it must be a careful choice.

- **Manual Wheelchairs**

Lightweight manual wheelchairs are often used by people who retain **trunk stability and the ability to self-position** themselves in the chair. These chairs are light to push and may be easily transported by car. Because ALS is a degenerative disease, it may be more cost-effective to rent or borrow this type of chair, leaving funds available for other equipment.

- **Power Wheelchairs**

Power wheelchairs allow a person to remain mobile and independent for a longer period of time than manual wheelchairs do. A standard power chair (or scooter if muscles are relatively unimpaired) will increase your outdoor mobility while reducing overall fatigue. Because the progression of the disease will necessitate ongoing changes, borrowing or renting this type of chair may be more cost-effective than buying it. These chairs are extremely expensive.

As you weaken and more support and better positioning are needed, a scooter will no longer be a viable option. Wheelchairs with manual or electric tilt-and-recline functions should be considered.

Many people with ALS find breathing easier when in a reclining position. This type of chair positions the body in ways that use gravity, instead of working against it. The tilt feature allows gravity to pull the hips to the back of the chair to prevent a continual forward sliding movement, which is a common problem with a reclining chair. **The wheelchair should support your back and head.** Specialty backs provide pressure relief and lateral support, which improves stability and positioning. Headrests are available in a variety of sizes and styles. **The tilt feature also relieves pressure** on the ischials and coccyx (tailbone), preventing pressure sores from forming.

No matter what type of chair is used, skin breakdown is more likely to occur if regular repositioning techniques are not used. **Cushions should be chosen for comfort and pressure relief, as many positions demanded by prolonged wheelchair use can cause skin breakdown.** If you can independently reposition every 30 minutes, a high-density foam cushion should be adequate to relieve pressure. As physical mobility decreases, more pressure-relieving cushions will be necessary. Many people with ALS report that gel cushions are uncomfortable, while air-filled cushions seem to provide more comfort and pressure relief. More than one type of cushion is usually required since no cushion will provide both comfort and relief all the time.

Wheelchairs can be controlled using many different methods. A therapist can help you determine what method best meets your needs.

Lifts and Stair Glides

This type of equipment can be expensive and requires much consideration and pre-planning to determine what is the best solution for your setting home and longer-term situation. The types of lifting equipment to consider include stair glides, portable lifts, ceiling-track lifts and wheelchair lifts. **It is best to get a trained professional to assess your residence and situation when making your plans regarding lifting devices.**

- **Chair Lift**

People with weakened legs will find it difficult to get up out of chairs. Higher chairs with arms to push on should make it easier. Ultimately, an **automated easy-lift chair** may be required. These are motorized chairs that have a switch enabling the person to adjust the incline, and to raise the seat to lift themselves out of a sitting position and into a standing position. More expensive models come with heaters, a massage device and other features.

- **Ceiling Lifts**

Ceiling lifts are usually installed to move a person between the bedroom, bathroom and living areas. In this case a rail is installed in the ceiling, and the person is lifted in a sling, similar to the sling used on a portable lift. There are also turntable tracks that allow the running tracks to intersect or cross. Installing a ceiling lift system is a major project that requires professional advice and installation.

- **Portable Lifts**

Portable hoists or lifts, which use a sling to lift a person up, can be rolled around on one floor level. A common type of sling lift is the **Hoyer Lift**. Most of these kinds of lifts are light, can be broken down into two pieces, and easily moved to a different level, or put in a car. The minimum door size for moving a person through in a portable lift is about 26". Using a portable lift requires training, during which both you and your caregiver should be lifted, so that the caregiver can understand what you are experiencing.

- **Stair Glides**

Most stair glides use a track that is fastened securely to the wall side of a stairway. An automated chair can then be moved up and down the track. Stair glides can go on curving stairs, and even around corners. However, there must be adequate room and help at the top and at the bottom of the stairway, for assisting you on and off the stair glide chair. **Stair glides should only be installed by a professional familiar with the safety aspects of this equipment.**

- **Wheelchair Lifts**

Portable wheelchair lifts are the most economical solution, and can be used on various stairways. Built-in wheelchair lifts are often used when it is not practical to have a wheelchair ramp from an outside door to ground level. Again, **installing a wheelchair lift system is a major project which requires advice and installation by a professional who is familiar with the safety aspects of wheelchair lifts.** (An amateur installation of a wheelchair lift was once responsible for the death of a small child in Ontario.)

Bathroom Equipment

There are several devices to help people with disabilities in the bathroom. **Raised toilet seats** or **commode chairs** are a standard requirement for those with weakened leg muscles. Raised seats can range from home-made and low-cost seats to more expensive models. Another option is to have a plumber raise the level of the toilet by putting it on a low platform.

Attachments to give a standard toilet a **bidet** function are also available. Some people with ALS who have little lower body disability, but have problems with their arms and hands find using a bidet enables them to retain total independence with toileting.

When it comes to bathing, there are a number of assistive devices. There are several models of **bath tub seats** and **lifting equipment** that can assist you in sliding or rotating over the tub for showering.

Retail stores that handle equipment for the disabled often have a variety of bathroom aids on display. Check with your occupational therapist or other healthcare professional for their assistance in choosing these aids.

Beds and Mattresses

A common problem during the advanced stages of ALS is being unable to roll over in bed. Lying in one position becomes intolerably painful, or requires a caregiver to move you every few hours. **Satin sheets** can make turning easier. There are now specially segmented, **air filled mattresses**, (a PVC- Poly Vinyl Chloride-bed) which are attached to an electronic pumping device which continually changes the air levels in different cells

in the mattress, thereby relieving pressure during the night.

A bed with an incline feature can be helpful with breathing and comfort. Devices from **foam wedges** to fully automatic hospital beds are used. **Hospital beds** are often advantageous when lifts (e.g., Hoyer lift) are used because of the extra clearance required underneath.

Augmentative and Alternative Communication (AAC) Equipment

There are many different types of assistive devices available to help the speaking impaired communicate. These devices range from **communication letter boards** based on tracing the person's eye movements (low-tech), to **speaking valves** for those on a respirator, to small **hand held electronic speaking devices**, to **computer based systems with sophisticated software and speech synthesizers** (high-tech). Your choice will depend on your budget and/or other funding available, your specific needs, and your adaptability to new ways of communicating.

Keep in mind that assistive technology is continually improving. A representative from your local ALS Society should be able to help you identify equipment resources in your community and may have some types of AAC equipment for loan.

Acquiring AAC equipment is only half of the equation. **It is critical that you are properly assessed by an assistive technology expert, who may be a speech language pathologist or occupational therapist and trained how to use the devices.** It is important for your family caregivers to also be trained.

See **Section 4**,
Sub-section *Adapting to Changes in Speech and Maintaining Communication.*

Tube Feeding Equipment

Tube feeding normally requires two pieces of equipment:

1. **Container** with a tube and clamp for the liquid food
2. **Pole** to hang the container on

A pump may also be used instead of a clamp to regulate the liquid flow for a short period (2 to 4 weeks) while you get used to tube feeding. A special syringe can also be used to inject liquid through a tube, or to check if your stomach has too much liquid.

- **Containers**

Feed containers come in the form of plastic bags or bottles with a tube attached. Some containers have a stretchable section, which is required if a pump is being used. These containers need to be thoroughly cleaned every day and replaced with a new container approximately every seven days.

When choosing a container you will consider various factors:

- **Format:** A plastic bag or plastic bottle
- **Capacity:** Common sizes range from 500 to 1000 ml
- **Tube:** You may need the type of tube that works with a pump

- **Poles**

It may be practical to rig up your own method for hanging the feeding container. One way is to use a pole held up by a Christmas tree stand. If you wish to purchase a pole, the commonly available models are priced from under \$100 to over \$400. Most models are extendible, have casters, and two or four hooks.

When choosing an IV pole, you should consider the following options:

- **More expensive models** tend to be more sturdy and stable
- **Floor models** or other models that attach to a table, chair or a bed
- **Pocket model** available for travelling. It has fittings that will attach to a variety of surfaces, including a suction cup for windows, a magnet for metal surfaces, an eye and two hooks, one of which is for the food container.

See **Section 4**,
Sub-section *Adapting to Swallowing Problems and
Maintaining Good Nutrition.*

HOME MODIFICATIONS

Listed below are some considerations to keep in mind when making home modifications to accommodate wheelchair users:

- **Rearrange furniture** to make wheelchair access and movement easier
- **Install ramps** (12:1 in slope) and guardrails or lifts in place of stairs
- **Doorways** should be at least 76 cm (30 in.) wide. They can be widened by removing the door and hinges, or removing inner moldings and installing a sliding door or curtain.
- **Hallways** should be 92 cm (36 in.) wide to accommodate the chair's turning radius. A cleared turning space of 1.6m² (five sq. ft.) is necessary for safe and easy turns
- **Space under sinks and counters** to accommodate wheelchairs can be made by removing cupboards and exposed pipes must be insulated
- **Thick carpets can hinder** the movement of a wheelchair; Berber carpet, wood, laminate, or linoleum are good choices for wheelchair users
- **Shower renovations** for wheelchair access

Some families will choose to make extensive home modifications while others will not. **The financial cost is a major consideration.** However, it is important to note that while it may seem like a good idea to make modifications well in advance of needing them, given your expectations of the disease process, keep in mind ALS is very individual. Your disease may progress slowly and spending money on a roll-in shower now, may not be something you need for several years if ever. The more expensive the modification, the more consultation you will need with your occupational therapist. **It is also a good idea to talk to other families who have been affected by ALS to share their thoughts, opinions, and experiences.**

Final Note on Assistive Equipment...

The most important messages to remember when it comes to assistive equipment are to:

- Think about both short- and long-term needs
- Consult with appropriate healthcare professionals about your equipment options and how to use what you select
- Learn from others who have come up with creative, simple solutions
- Seek information on financial assistance and equipment loan programs to minimize your out of pocket expenses

LIVING WITH ALS CAN BE EXPENSIVE

Having ALS can be very expensive, whether one lives at home or in an institution. Institutional care imposes a heavy burden on taxpayers. While home care is much less costly, the expense falls principally on the family, and often becomes exorbitant in terms of their ability to pay. This is true even though various agencies and groups provide some forms of help. **The assistance provided by the ALS Society becomes, for many people, a critical factor in remaining in the home setting.**

A person with ALS may need some form of assistive device at the time of diagnosis, and in almost all cases, will proceed through a bewildering variety of equipment: cane, walker, manual and power wheel chairs, transfer seat, ceiling lift, stair glide, porch lift, hands-free toilet, ventilator... The list goes on and on. It may be that having no other condition requires so much equipment or care.

In addition to equipment, persons with ALS take many medications for symptom management. Depending on whether or not you have a generous drug benefit plan, this can get expensive.

Home care assistance and respite care enabling your family caregiver a break also comes at a financial cost to the family.

Some families feel it is important to purchase a wheelchair accessible van or make substantial modifications to their home to help the person with ALS maintain independence.

Obviously, these choices come with rather hefty price tags. Funding assistance for some of the associated costs of ALS may be available through government and other private programs. Talk to your social worker (or case manager), or contact your local ALS Society office for more information and referrals. In many cases, your local ALS Society will be able to help you directly with equipment through an *Equipment Loan Program*.

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MANAGING YOUR EMPLOYER

When you have first been diagnosed with ALS, there are many things to consider. One is when and what to tell your employer. This is very much a personal decision depending on your condition and personality, as well as your employment situation.

Checking out the **benefits** in your employer's group insurance is certainly a first step, though at this stage you may not yet be aware of the full range of your potential needs. Be careful about doing anything that might restrict your access to group benefits. The **pension plan** may be another area worth checking into. **All of this can be done before telling your employer you have ALS.**

When Do You Tell Your Employer?

Here are some things to consider:

- Can you hide your symptoms from your employer?
- Are you working on a project that you want to finish, before telling your employer?
- Should you tell your employer so that they have adequate time to plan how to accommodate you in the work place or plan for your replacement? (In many provinces it is a legal requirement to "accommodate" a person with a disability such as ALS.)

Do not sign anything regarding your termination without first getting legal advice. Legal termination minimums are usually linked to years of service.

What Protections Do You Have Under Provincial Labour Legislation?

Consider getting advice from a lawyer who specializes in human rights, provincial employment standards and labour legislation.

INSURANCE ISSUES

Understanding Your Health Insurance Coverage

In order to maximize the benefits of your insurance plan, it is important for you to keep a record of the name, phone number, identification, and group number of your insurance plan. You should also note who the subscriber is (you or your spouse), his/her date of

birth, and social insurance number. Identify any other insurance benefits you may have, such as a separate prescription benefit card or a long-term-care policy.

If you have more than one insurance plan, determine which policy is primary and which is secondary; confusion with this matter can result in billing errors. Your insurance company customer service representative can be helpful in identifying which plan is to be billed first. Also keep up to date on your insurance; if your coverage changes in any way, notify all your doctors, medical suppliers, and pharmacists immediately so that the next time you access their services, you will have helped to prevent billing errors.

Contact your insurance company directly, and ask specific questions about your benefits. Always note the date and the person who provided the information. Remember, having your benefits described over the phone does not guarantee coverage. Your doctor may be asked to write a letter of medical necessity and complete specific forms to verify your medical condition and eligibility for whatever item or service is being requested.

Ask if your policy offers case management. Case managers either work for the insurance company or are contracted by them to monitor and advocate for clients whose costs are high or who have complicated needs. They are often helpful in gaining access to little-known insurance benefits, and reducing your overall out-of-pocket expenses. Case management can be activated at various stages of your illness, depending on how your insurance company has defined the benefit. Having direct communication with your insurance carrier will help you to gain an overview of your policy and remove the mystery of how your plan works.

Use the following guide when contacting your insurance company. Be sure to have all of the specific information about your insurance plan in front of you before calling. Do not hesitate to ask what certain words or terms mean.

Group insurance can be very important to a person with ALS. If you have group insurance through your employer, you may need to seek advice from a lawyer or someone else you can trust to ensure that this insurance coverage continues to be effective when you are unable to continue working. Some employers attempt to terminate employees who become disabled. They do so to cancel the disabled employee's group insurance to keep company insurance premium costs down. There are also many cases where persons with disabilities have had to hire lawyers to force an insurance company to pay the benefits described in their group insurance plan, particularly loss of income benefits. If you require legal advice for this purpose find a lawyer who has experience in this area.

Driving and Property Insurance

If you don't report your disability to your insurance company, your car insurance coverage may not be valid. Driving can sometimes require fast foot and hand reactions to avoid an accident. Ask at your clinic about a driving test service that will certify your current abilities for insurance purposes. Also, you may need additional property insurance regarding assistive equipment you have or renovations that have been done in your residence to accommodate your condition.

Life Insurance: Living Benefits

Most life insurance companies offer a "living benefit" feature to people with a terminal disease enabling them to get a portion of their life insurance paid to them in advance, during the years before their death. If you wish to benefit from such a plan, calculate how much the benefit is worth, relative to the resulting decrease in amount of your life insurance. If you do not understand these calculations, get advice from an independent person you can trust. For more information on living benefits see the following pages.

Guide to What You Need to Know About Your Health Insurance Policy

General Questions

- Is there an annual deductible?
- Is there an annual out-of-pocket expense limit or maximum? If I meet my limit, does my coverage increase and to what extent?
- Do I have a major medical plan? Is there an annual or lifetime maximum?
- Do I need to complete any claim forms?
- Am I subject to preexisting condition regulations?
- Do I have a yearly or lifetime maximum limit?

Durable Medical Equipment (DME) Questions

- Does my plan cover DME? What about ventilator coverage; is it under respiratory equipment or DME?
- What is the percentage of my coverage?
- Is there a preferred provider I must see?
- Is pre-authorization or a medical review required?

Prescription Questions

- Does my plan cover prescription drugs? What are the terms of this coverage, and is coverage different based on using brand-name versus generic drugs?
- Is there a specific pharmacy/supplier network I must use?
- Are injectable medications covered under my plan?
- Is there a limit on the amount of prescription drugs I can go through on this plan?
- Is there coverage for all Health Protection Branch-approved drugs, or is coverage provided only for those listed on your formulary (a list of drugs that an insurance policy covers)?
- Does my plan offer a mail-order pharmacy option? How does this work?

Group Medical Insurance Questions

Examine your group insurance policy carefully and call your insurance provider for detailed information.

- What are you entitled to?
- Loss of income support: How much and for how long?
- Prescription Drugs: What drugs are covered? What is the maximum? What is deductible?
- Alternative Therapies: Are any, such as massage, covered? Any limitations?
- Home Care: How much and how long?
- Assistive Equipment: What type and what limitations?
- Hospital: Private or semi-private coverage?
- Are there any other benefits?

Home Health Questions

- Does my plan have home health coverage? How do I access this?
- Is there a preferred home health care agency I must use?
- Is there private-duty nursing coverage at home? Describe this benefit.
- Does my plan offer case management? At what point does case management get involved and for how long?

- **How do living benefits work?**

Policies and procedures for living benefits vary slightly, depending on the insurance company. Typically, to receive living benefits an application must be made to the insurance company together with a medical opinion that **the policy holder is in the terminal stages of an illness, and has approximately 24 months to live.**

The insurance company will then check to make sure the proceeds of the insurance have not been assigned to pay off a loan or debt or left irrevocably to someone else who might sue for the full proceeds once the policy holder has died. Once these two conditions are met, the insurance company will then pay a percentage of the value of the policy - ranging from one-third to half - as a living benefit. Again, the details vary depending on the company. Some set a maximum - such as \$50,000 - for living benefits. Some charge interest, which is usually paid out of the policy. Some make special arrangements with the person to ensure that the regular premium payments are made and the insurance policy stays in force.

- **What happens to the money left in the policy?**

When the policy-holder dies, the remainder will be paid to his or her estate or designated beneficiary, less any interest charges on the money paid out as living benefits.

- **Are living benefits available on all life insurance policies?**

Living benefits are usually available on individual life insurance policies but they are **not always available on group life insurance plans.** Practices vary, so be sure to ask specifically about your policy.

- **What should I do if I am interested in getting living benefits?**

Before you decide to apply for living benefits, **talk to a lawyer or financial advisor about all the financial implications.** At the present time, the federal government is not taxing living benefits, but anyone receiving a lump-sum living benefit payment will no longer be eligible for social assistance benefits. Talk to someone who can look at your entire financial situation and

advise you about the best course of action.

If you decide to apply for living benefits, either you or your lawyer should talk to your insurance agent. Ask about the availability of living benefits under your policy. Because this is a relatively new program in most insurance companies, some agents and brokers may not be aware of it. If you are having trouble getting information about your policy or getting living benefits, contact the help line at the Canadian Life and Health Insurance Association: in Toronto 416-777-2344; outside Toronto 1-800-268-8099.

Caution: There are organizations that buy life insurance policies from people who are terminally ill. In return for a certain amount of cash now, usually substantially less than the death claim value of the policy, the person with a life insurance policy names the organization as beneficiary and gives up all rights under the policy. When the person dies, the organization will receive the full value of the policy.

In Ontario at the present time, it is illegal for any organization not licenced to sell insurance policies to do so. While there are no such organizations operating in Ontario, many U.S. firms actively try to recruit Canadian policy-holders. Talk to your lawyer or the Canadian Life and Health Insurance Association before becoming involved with these organizations.

PENSION PLANS

Group Pension Plans

If you are in a company or other group pension plan, find out what will happen as a result of an ALS diagnosis. To ensure that you get your full entitlement, have these questions answered by someone you trust.

- Can you receive some of your pension before age 65, if you are disabled, and unable to work?
- Will payments be made to your spouse if you are no longer alive?

If problems occur, you may need advice from a lawyer with experience with group pension plans.

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To minimize tax on death, you may want to make sure that your spouse is the beneficiary of your pension plan; otherwise your pension plan's value will be part of your estate, and subject to probate fees.

Persons with ALS may qualify for a CPP disability pension. To be eligible for a Canada Pension Plan Disability Pension, you must:

- Be between the ages of 18 and 66
- Have contributed to the CPP for a minimum qualifying period calculated as follows:
 - If you have only two calendar years in your contributory period, you must have contributed in both years.
 - If you have more than two years in your contributory period, then contributions must have been made in two of the last three years, or five of the last 10 years of the contributory period.

When you are already receiving a CPP retirement benefit and become disabled between 60 and 65 years of age, you must have become disabled before, or within six months after, the effective date of your retirement pension. Apply in writing. **(Applications can be obtained from your nearest federal government office.)** A Canada Pension Plan Disability Pension is payable from the fourth month after you are deemed to have become disabled. You may receive up to a maximum of 12 months of retroactive payments. Like most pensions, your CPP Disability Pension is considered taxable, subject to your total income.

Things that Can Change Eligibility for a CPP Disability Pension:

Those receiving a CPP Disability Pension must notify the Canada Pension Plan of any changes that might affect their continuing eligibility for benefits. This includes:

- An improvement in your medical condition
- A return to full, part-time, volunteer or trial period of work
- Attendance at school or university; trade or technical training
- Any vocational rehabilitation

TAX ISSUES

Disability Tax Credit

Form T2201, Canada Revenue Agency (CRA) *Disability Tax Credit Certificate*, has a section that must be completed and signed by your doctor and sent in with your income tax return.

Minimize Tax Through Income Splitting

Income splitting reduces a family's total income tax by dividing income among various family members, using up the low tax rates of family members who have little other income. This has to be done in ways that will "attribute" less income to the higher income earner(s) in the family, according to CRA, such as the following:

- Estate freezing
- Transfer property at fair market value
- Lend or gift assets to generate business income, or income on income
- Gift funds to child turning 17
- Earn capital gains for children
- Reasonable salaries (for example, person with ALS paying spouse as a caregiver is a deductible expense to the person with ALS)
- High taxpayer pays all household expenses
- Deposit child tax credits in child's bank account
- Contribute to spousal RRSP
- Assign one half of CPP benefits to spouse

And there are others. If substantial funds are involved, or even if funds are minimal, see a professional tax expert who specializes in disability issues.

Estate Freezing

The purpose of estate freezing is to **minimize the taxes due on death**. Assuming assets will be at a taxable level, taxes can be minimized by a financial plan that is structured so that future profits from your assets will go to someone else in a lower tax bracket, such as your children. Consider getting advice from a professional accountant regarding estate freezing.

Tax on Deemed Disposition of Assets on Death

At the date of death, the CRA requires one tax return for income earned to the date during the year and to account for the tax that is applicable on all of the increased values of the deceased person's properties and other assets. There are certain tax-free "roll over" provisions for property left to a spouse, or spousal trust. Also, dividend-paying shares may incur a double tax unless appropriate action is taken. **Be sure to seek advice from a professional accountant on these matters.**

Probate Fees

A Certificate of Appointment of Estate Trustee with a Will validates a will. Upon issuing this certificate, the Estate Court charges a tax rate that varies from province to province. Probate fees are reduced by reducing the amount of the estate, such as making life insurance payable to a spouse instead of to the estate.

U.S. Estate Tax

If you were born in the U.S. and own property in the U.S., or own U.S. securities registered in your name, your beneficiaries may be in for an unpleasant surprise.

Even if you have been a Canadian citizen for most of your life, when the U.S. Internal Revenue Service (IRS) learns of a person's death, they will apply an estate tax on the total estate, regardless of what countries the assets are in. Assets will then be held by the IRS until this tax is paid. There are easy ways to avoid this tax. If this could happen to you, **see a professional accountant who is familiar with U.S. estate taxes.**

Tax-Effective Wills

There are numerous estate planning alternatives, including naming a spouse as beneficiary to all life insurance and pension plans. Also multiple testamentary trusts, an exclusive spousal trust and trustee powers to authorize actions to minimize tax are other techniques. Mentioning these plans in a will helps to ensure that your plan will be carried out according to your wishes. **Bequests to a voluntary organization can also reduce estate tax.** Again, if substantial amounts are involved, it is probably wise to consult with a professional estate planning accountant or estate planning lawyer.

LIVING WILLS AND POWERS OF ATTORNEY

What is a Living Will?

A living will, sometimes called an advance directive, is a document that states your written instructions about your wishes for future health or personal care. For example, some people include their choices regarding "Do Not Resuscitate" orders (DNRs), being connected to machines to stay alive, having surgery, and donating organs. Personal care includes choices regarding shelter, nutrition, safety, hygiene, comfort, etc.

A living will only takes effect when you are no longer able to understand treatment choices and consequences, or too ill to communicate. For someone with ALS, in the event of an emergency, like respiratory failure, and the person does not have the capability to communicate their wishes, a living will would take effect.

Depending on the province in which you live, legally recognized living wills may be called "health care directives," "advance health care directives," "representation agreements," "mandates," "authorizations," "personal directives," and "powers of attorney for personal care."

There are two parts to a living will: a *proxy directive* that specifies who you want to make decisions for you on your behalf if you cannot (see Power of Attorney for Health Care) and an *instruction directive* that specifies what health care or other personal care choices you would want your proxy to make in particular situations.

A "Values History" is something you may also want to include in a living will. An explanation of your personal beliefs and values may make it easier for decision-makers to understand and follow your wishes.

What Is a Power of Attorney?

A Power of Attorney is a document that gives legally recognized decision-making power on your behalf to whomever you have appointed to do so in the event you are incapable due to limited mental ability or being too ill to communicate. You may choose one or more persons to act as your substitute, or "proxy." If you select more than one person,

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you must indicate whether any named individual can make decisions for you alone, or whether they must make decisions as a group.

There are **two kinds** of Power of Attorney documents: 1) for property, and 2) for personal care.

1. Continuing (Enduring) Power of Attorney For Property

A Continuing Power of Attorney for property (including managing your financial affairs) **will help to protect your assets from being taken over by the provincial public trustee.** If that happens, your family will have an easier time getting back control of your assets if they have your Continuing Power of Attorney.

2. Power of Attorney for Health Care (PAHC)

A Power of Attorney for health care may be referred to by various terms from province to province (e.g., Power of Attorney for Personal Care). No matter what it is called, its function is the same. It is a legal document **used to name the person(s) to whom you want to give legal authority to make decisions for you in the event you cannot regarding medical treatment, according to your living will** and not on the basis of someone else's assumptions. Should the specifics of a particular situation not be covered in the living will, your appointed "proxy" can act on your behalf.

It is exceedingly important that **when appointing your substitute decision-maker**, the person(s) you select are known well to you and can be trusted to make decisions on your behalf as previously instructed by you. You must ensure that they are aware of your wishes so that they will follow them unless it is impossible to do so. Your **Power of Attorney for Health Care** (or whatever it is named in your province) should be kept in a safe place, and easily accessible to the person(s) you have appointed. You may want to carry a wallet card that names your Power of Attorney(s) and includes their contact information.

Your living will can be either a separate document, or you can enter your personal and healthcare wishes on your PAHC form recognized in your province. A separate document may give you more flexibility to be very detailed.

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Why Are these Documents Important?

A living will and PAHC are important because they:

- Promote self-determination and if followed they satisfy the wishes of the declarant
- Reduce stress associated with difficult decisions often passed on to immediate family and friends
- Release medical practitioners from legal and ethical repercussions.

If there is no living will, there is no telling how close to your actual wishes the care you receive will be. **Having a living will is particularly important for someone with ALS in the event of a respiratory crisis.** You need to make it abundantly clear in advance whether or not you would want a tracheostomy ventilator to save your life.

Do I Need a Lawyer to Complete the Documents?

Not necessarily, but it may be helpful to have a lawyer with expertise in this area consult with you to make sure it is legally valid in your province.

When Should the Documents Be Completed and Given to Whom?

It can be very difficult and stressful to complete a living will and appoint a healthcare proxy when you are in the latter stages of a disease. Therefore, it makes sense to **initiate the process when you are relatively healthy.**

Once you have completed your documents, provide your **doctor and clergy** copies as well as your **PAHC appointee(s)** and other **members of your family.** Both family and healthcare providers need to know your wishes to increase the odds that they are respected in the event you are unable to communicate them for yourself.

Can a Living Will or Power of Attorney Be Changed?

Yes. Advance directives should be viewed as dynamic. However, if changes are made, you must destroy all pre-existing versions and distribute replacements to the people who were given earlier ones. As a person with ALS, you will experience many changes in health and functional status, and as such, your perceived quality of

life will also change. Therefore, what you decide you do not want today, may change a year from now. **It is advisable to revisit your living will every six months or so.**

Legal Recognition

Legal recognition of living wills and power of attorney documents **varies from province to province**. In provinces with specific legislation, people may be legally required to follow your living will. The Canadian Medical Association has endorsed a policy supporting living wills and most doctors favour them.

To understand the laws in your province talk to a lawyer. You may also contact or visit the web site of your province's Ministry of the Attorney General for more information.

Where Can I get Sample Advance Directive Documents?

Power of Attorney and living will kits are often available at no charge through your **provincial government** and can be downloaded on-line. Living will forms and kits can also be purchased at bookstores or via the internet. A sample living will form is available at no charge through the University of Toronto Joint Centre for Bioethics. You may view and print the entire document which provides detailed background about living wills, the legalities of living wills province by province, information about healthcare decisions, and personal care decisions at

www.utoronto.ca/jcb/home. This document was prepared in 2002. For up-to-date Information on the status of living wills in Canada, consult with a lawyer. A sample healthcare directive created specifically for palliative care patients is available on-line at www.palliative.info. An advance care directive form designed at the University of Washington specifically for patients with ALS (focusing on feeding tubes and invasive mechanical ventilation choices) is found in the following journal article:

Benditt, Smith, and Tonelli (2001). Issues and Opinions: Advance Care Planning in ALS. *Muscle and Nerve*, 24: 1706-1709.

To request a copy of this article please contact the director of services and education , ALS Society of Canada at 1-800-267-4257 x230.

LAST WILL AND TESTAMENT

Why is a Will Necessary?

An estate of a deceased person will not normally come under the control of the Public Guardian and Trustee (formerly the Public Trustee) if there are family members who are entitled to the estate. The family members who are entitled to receive the estate are also entitled to be appointed administrator (or Estate Trustee).

If the family members are not entitled to be named as administrators, or do not wish to take on the administration, it is possible for them to name the person who will take on this responsibility. However, having a will is a much better option.

Planning A Will

Planning your will can minimize the additional income taxes, and probate fees payable on your death. Also, planning can minimize taxes payable by your beneficiaries on future income.

To plan a will you will need to:

- Make a list of all assets and liabilities including additional taxes due on your death
- Determine who your beneficiaries will be, such as spouse, children, other relatives, friends, charities, etc.
- Consider tax reduction strategies, such as donations to charities
- Choose your estate trustee, such as your spouse or trusted friend. A lawyer, accountant or trust company could be considered for more complex situations

Using Your Will to Make That Special Gift

Many people choose to make provision in their will to contribute a special bequest to a cause close to their heart. Bequests allow people to name a charity of their choice as a beneficiary and to make a financial contribution from their estate to the charity that is often far greater than would have been possible during their lifetime. You may wish to consider the ALS Society in your province, or the ALS Society of Canada, as a beneficiary for a fixed donation or a percentage of your estate.

Final Note on Legal and Financial Considerations...

On top of coping with the day-to-day challenges of living with a progressive illness like ALS, financial and legal planning can seem overwhelming. However, it is very important to consider these aspects of life for yourself as well as your loved ones. Perhaps asking a family member or friend to help get you organized or do research on your health benefits and legal rights can make it more manageable.

PRINT MATERIALS

ALS Medical Textbooks

Amyotrophic Lateral Sclerosis

By Hiroshi Mitsumoto, M.D, et al
 Oxford University Press
 2001 Evans Road
 Gary, NC 27513
 (800) 451-7556
 ISBN: 0803602693

Amyotrophic Lateral Sclerosis: Diagnosis and Management for the Clinician

Edited by: Jerry M. Belsh, MD and Philip L. Schiffman, MD
 Futura Publishing Company, Inc.
 135 Bedford Road
 P.O. Box 418
 Armonk, NY 10504-0418
 (914) 273-1014
 ISBN: 0879936282

Motor Neuron Disorders

Edited by:
 Pamela J. Shaw, MD
 Michael J. Strong, MD
 Butterworth-Heinemann/Elsevier, Inc.
 Independence Square West
 Philadelphia, PA 19106
 (215) 238-2239
 ISBN: 0750674423

Palliative Care in Amyotrophic Lateral Sclerosis: Motor Neuron Disease

By David Oliver, Gian Domenico Borasio and Declan Walsh
 Oxford University Press
 ISBN 0192637667

ALS Management Guides and Coping with Chronic Illness

Amyotrophic Lateral Sclerosis (NEW!)

By Robert G. Miller, MD, Deborah Gelinis, MD, & Patty O'Connor, RN.
 This book is one of the first in a series sponsored by the American Academy of Neurology design to assist people with neurologic diseases and their families. The

authors have included chapters about the disease itself, specific symptoms and how they can be managed, how multidisciplinary centres work, how voluntary health agencies help, how computers can help and more.
 Demos Medical Publishing, Inc.
 386 Park Avenue South
 New York, NY 10016
 ISBN: 1932603069

Amyotrophic Lateral Sclerosis: A Guide For Patients and Families, 2nd Edition

By Hiroshi Mitsumoto, MD & Theodore L, Munstat, MD
 This comprehensive guide covers every aspect of the management of ALS. Beginning with discussions of its clinical features of the disease, diagnosis, and an overview of symptom management, major sections deal with medical and rehabilitative management, living with ALS, managing advanced disease and end-of-life issues, and resources that can provide support and assistance.
 Demos Medical Publishing, Inc.
 386 Park Avenue South
 New York, NY 10016
 ISBN:1888799285
www.demosmedpub.com

ALS: Maintaining Mobility

This is a guide to physical therapy and occupational therapy. It illustrates techniques for energy conservation, managing architectural barriers and employing adaptive devices.
 ALS Neurosensory Center
 6501 Fannin Street, Room B310
 Houston, TX 77030

Communication and Swallowing Solutions for the ALS/MND Community

Edited by Marta S. Kazandjian, SLP, CCC
 This invaluable resource manual enables the person with ALS/MND, caregivers and family to make informed decisions to best manage communication and swallowing difficulties as they arise and illustrates how these solutions can be used to support and resolve the individual's needs.
 Singular Publishing Corp.
 San Diego, CA
 (800) 347-7707
 ISBN: 1565938089

Les Turner ALS Foundation Resource Guide

The third edition of the Les Turner ALS Foundation Resource Guide provides information and resources for ALS patients, family caregivers and health professionals. Assembled by the team members of the Lois Insolia ALS Center at Northwestern University's Feinberg School of Medicine, the Resource Guide is available online for viewing, downloading and printing with Adobe Acrobat or you may order the print version through the Foundation.

To order a printed and bound copy of the Resource Guide, please contact Claire Owen, Director of Patient Services at cowen@lesturner.org (847) 679-3311.

Life on Wheels-For the Active Wheelchair User

By Gary Karp

This book offers practical ways to adapt and optimize the quality of your life. It covers subjects such as skin care, bowel and bladder care, sexuality, home access, maintaining a wheelchair and dealing with insurance problems.

O'Reilly and Associates, Inc.

LaVegne, TN

(800) 998-9938

ISBN: 1565922530

Living with ALS Manuals

Six manuals on managing ALS to assist patients, their families, and health care practitioners:

1. What's it all About?
2. Coping with Change
3. Managing Your Symptoms and Treatment
4. Functioning When Your Mobility is Affected
5. Adjusting to Swallowing and Speaking Difficulties
6. Adapting to Breathing Changes

The ALS Association

27001 Agoura Road, Suite 150

Calabasas Hills, CA 91301-5104

(800) 782-4747

(manuals are free of charge to ALS families and healthcare professionals caring for persons with ALS)

Meeting the Challenge-Living with Chronic Illness

By Audrey Kron, MA, CGP

Discounts on orders of 10 or more copies are available through the publisher.

Center for Coping with Chronic Illness

West Bloomfield, MI

(248) 626-6960

Realities in Coping with Progressive Neuromuscular Diseases

This book brings together 51 eminent authorities on ALS focusing on a variety of different coping strategies to offer to patients and families, as well as health professionals

Charles Press Publishers, Inc.

P.O. Box 15715

Philadelphia, PA 19103

ISBN: 0914783203

We Are Not Alone: Learning to Live with Chronic Illness

By Sefra Kobrin Pitzele

This book offers practical strategies and inspiration to people with chronic illness; not specific to ALS.

Workman Publishing

708 Broadway

New York, NY 10003

(212) 254-5900

ISBN: 0894801392

Cook Books**The Dysphagia Cookbook: Great Tasting and Nutritious Recipes for People with Swallowing Difficulties**

By Elaine Achilles, Ed.D.

Cumberland House Publishing

431 Harding Industrial Drive

Nashville, TN 37211

ISBN: 1581823487

Easy to Swallow, Easy to Chew Cookbook

By Donna L Weihofen, RD, Joanne Robinson, PhD, and Paula Sullivan, MS

This book offers over 150 tasty and nutritious recipes with simple instruction for tailoring food textures from very easy-to-chew to soft and smooth. All recipes contain nutritional information per serving.

Wiley Publishers

ISBN: 0471200743

Meals for Easy Swallowing

Includes a collection of recipes, swallowing tips, helpful hints for increasing calories and sample menus developed by ALS patients and their families.

ALS Clinic

6501 Fannin, Room B 310

Houston, TX 77030

Non-Chew Cookbook

By J. Randy Wilson

A discounted price is available through the publisher on orders of 10 or more copies.

Wilson Publishing, Inc.
Glenwood Springs, CO
(800) 843-2409

Pot Holders and Love Handles (cookbook)

By Lynda Cabela (deceased person with ALS)

Cabelas Furniture Store
602 Second Street
Cappell, NE 69129

Caregiver Books and Guides**Caregiving: The Spiritual Journey of Love, Loss and Renewal**

By Beth Witrogen McLeod

This book is written by a journalist with plenty of experience with family caregiving. Beth was a caregiver for both parents, one with cancer and the other ALS.

Wiley and Sons Publishing
ISBN: 0471254088

The Caregiver Survival Series

By James R. Sherman, PhD

In this series of booklets are the following titles:

Positive Caregiver Attitudes

ISBN: 0935538186

The Complete Bedside Companion-No-Nonsense Advice on Caring for the Seriously Ill

By Roger McFarlane and Philip Bashe

This comprehensive and compassionate handbook is an excellent guide for home caregiving to the seriously sick and dying. It includes field-tested practical advice and support for all phases of illness, from the onset of symptoms through terminal phases, and necessary steps after death. This book is recommended both for home caregivers and as a reference book for public and medical libraries.

Simon and Schuster
New York, NY
ISBN: 0684843196

The Caregiver Survival Series

By James R. Sherman, PhD

This series includes the following titles:

Positive Caregiver Attitudes

ISBN: 0935538186

The Magic of Humor in Caregiving

ISBN: 0935538194

Coping with Caregiver Worries

ISBN: 0935538208

Creative Caregiving

ISBN: 0935538178

Preventing Caregiver Burnout

ISBN: 093553816X

Pathway Books

Golden Valley, MN

(612) 377-1521

The Courage To Laugh: Humor, Hope, and Healing In The Face Of Death And Dying

By Allen Klein

This remarkable book is a poignant and inspirational reminder of the life-affirming nature of the human spirit, even under the most difficult circumstances. In the words of cartoon character Roger Rabbit, "Sometimes a laugh is the only weapon we have." Klein, uses a simple and approachable format to share stories of how people faced with death and illness use laughter for many purposes. It can serve as a defense mechanism, a tool for encouragement, a bonding between those in difficult situations, and a reminder that life goes on despite the worst. At its best it is a tool for coping, communicating, and connecting.

Jeremy P. Tarcher/Putnam

ISBN: 0874779294

The Comfort of Home: An Illustrated Step-by-Step Guide for Caregivers, 2nd Edition

By Maria M. Meyer with Paul Derr, RN

In this book, the authors provide an excellent guide to caregiving in the home. A chronological structure is used to define preparation for caregiving, the day-to-day expectations, and a listing of numerous resources to augment the content.

CareTrust Publications LLC

Portland, OR

ISBN: 0966476735

Family Caregiver's Guide: The Home Health Care Efficiency System that Really Works

By Joan Ellen Foyder

An easy step-by-step guide to caring for a patient at home. This book helps take the frustration out of home patient care by solving hundreds of everyday problems.

The Futuro Company
5801 Mariemont Avenue
Cincinnati, OH 45227
(513) 271-3782
ISBN: 0961739207

**The Four Things That Matter Most
A Book About Living (New!)**

By Ira Byock, MD

Four simple phrases -- "Please forgive me," "I forgive you," "Thank you," and "I love you" -- carry enormous power. In many ways, they contain the most powerful words in our language. These four phrases provide us with a clear path to emotional wellness; they guide us through the thickets of interpersonal difficulties to a conscious way of living that is full of integrity and grace. In *The Four Things That Matter Most*, Dr. Ira Byock, an international leader in palliative care, teaches us how to practice these life-affirming words in our day-to-day lives and helps us to forgive, appreciate, love, and celebrate one another more fully. Distilled from experiences of people who were forced by serious illness to face the impending end of their relationships, these stories convey practical wisdom that can help any person at any time say and do the things that really matter most in their own life. Dr. Byock asks, "Why wait until we or someone we love is dying to say the things that matter most? It would be great if this nugget of 'hospice wisdom' became part of public discourse and part of our culture."

Free Press/Simon and Schuster
(see www.fourthings.org)
ISBN: 0743249097

The Helper's Journey

By Dale G. Larson, PhD

This book is intended for volunteers, nurses, physicians, social workers, clergy, counselors and others who work on the front lines of caregiving, but may apply also to family caregivers. Most examples in the book are drawn from Dr. Larson's work in hospice, psychotherapy, and oncology.

In the Shadows: Living and Coping with a Loved One's Chronic Illness

By Dr. David Luterman

Research Press
2612 North Mattis Avenue
Champaign, IL 61821
ISBN 0878223444

Dr. Luterman is a professor of communication disorders who has given workshops on the impact of disabling disease on the patient's family. The book details his family's struggle with his wife's multiple sclerosis.

Jade Press
Box 822
Bedford, MA 01730
ISBN: 0964486202

Mainstay: For the Well Spouse of the Chronically Ill

By Maggie Strong

Bradford Books
ISBN: 0965717909

Share the Care: How to Organize a Group to Care for Someone Who Is Seriously Ill

By Cappy Capossela and Sheila Warnock

Drawing on the authors' personal experience in forming a caregiving network to care for a friend, this is a step-by-step guide to the group approach. This book offers valuable guidelines, compassionate suggestions, and a workbook that offers support to help free the person who is ill from worry and the primary caregiver from burnout.

Simon and Schuster
1230 Avenue of the Americas
New York, NY 10020
(Can also be ordered through www.sharethecare.org)
ISBN: 0684822367

What If It's Not Alzheimer's? A Caregiver's Guide to Dementia

Edited by Lisa Radin and Gary Radin

Foreword by Murray Grossman, M.D., Ed.D

Although today the public all too often associates dementia symptoms with Alzheimer's disease, the medical profession can now distinguish various types of "other" dementias that also undermine cognitive abilities, often with onset at a younger age. This book is the first comprehensive guide dealing with frontotemporal dementia (FTD), one of the largest groups of non-

Alzheimer's dementias. The contributors to this book are either specialists in their fields or have exceptional hands-on experience with FTD sufferers. This much-needed resource work, the first of its kind, provides a wealth of real and practical information to both healthcare professionals and caregivers of someone suffering from FTD.

Prometheus Books
ISBN: 1591020875

End-of-Life Books and Guides

Dying at Home: A Family Guide for Care Giving

By Andrea Sankar
ISBN: 0801842301

Dying Well

By Ira Byock, MD
Dr. Byock, one of America's leaders in palliative care medicine, shows us that much important emotional work can be accomplished in the final months, weeks, and even days of life. *Dying Well* brings us to the homes and bedsides of families with whom Dr. Byock has worked, telling stories of love and reconciliation in the face of tragedy, pain, and conflict. It provides a blueprint for families, showing them how to deal with doctors, how to talk to friends and relatives, and how to make the end of life as meaningful and precious as the beginning. Here is a book like no other on the subject: hopeful, clear-sighted, and life-changing.
Riverhead Books/Berkley Publishing Group
375 Hudson Street
New York, NY 10014
ISBN: 1573220515

Final Gifts: Understanding the Special Awareness, Needs, and Communications of the Dying

By Maggie Callanan and Patricia Kelley
This remarkable book by two hospice nurses shows how communication at end of life takes on special meaning. Touching case stories show how approaching death can give a clarity and importance to how we all relate to one another. The book affirms that you can live fully to the very end, and that the final gifts received by caregivers can outweigh the burdens they must carry. Practical suggestions on how to respond to the requests of the dying will be of value to anyone in a caregiving role.
Avon Books
ISBN: 0553378767

ALS. Three letters that change people's lives. FOREVER.

The Guide to End-Of-Life Care for Seniors

Edited by Rory Fisher, Margaret M. Ross, and Michael J. MacLean

This Guide is a collaborative effort between the Universities of Toronto and Ottawa. The purpose of the Guide is to support the end-of-life care of seniors that reflects a consensus of best practices in this area of healthcare and social service delivery. The Guide also emphasizes and encourages the collaborative nature of end-of-life care, supports personal health practices and decision-making, and strives to fulfill the adage that effective healthcare is able to "heal sometimes, relieve often and to comfort always."

University of Toronto, Interdepartmental Division of Geriatrics, Faculty of Medicine

University of Ottawa, School of Nursing, Faculty of Health Sciences

(416) 480-6068

www.rgp.toronto.on.ca/iddg/index.htm

The Guide to Recalling and Telling Your Life Story

This Guide is a beautiful workbook designed to help a person tell his or her life story. Page by page, it suggests topics--such as Family, Adult Life, Growing Older, and Reflections--and questions to consider. The questions can be helpful to the person sharing the memories, as well as provide other family members with a way to elicit stories and experiences. The Guide also includes a section in which to record values the person wishes to pass on to future generations. Published by Hospice Foundation of America
(800) 854-3402

Printable version available at

www.hospicefoundation.org

Handbook for Mortals : Guidance For People Facing Serious Illness

By Joanne Lynn, MD and Joan Harrold, MD
Foreword by Rosalynn Carter

A comprehensive and authoritative guide to end of life care, written for a general audience by a team of specialists that includes some of the world's leading authorities in the field. This readable and sensible book should be read by every family caring for a loved one who is seriously ill, and by every medical professional in a palliative care setting. The balance between content and presentation is extremely well done, making the book accessible to a general audience while still having

enough detail to be of value to medical professionals. Joanne Lynn, M.D. is President of Americans for Better Care of the Dying. Joan Harrold, M.D., is the Vice President, Medical Director of the Hospice of Lancaster County, Lancaster, Pennsylvania. ISBN: 0195116623

Last Touch: Preparing for a Parent's Death

By Marilyn R. Becker
New Harbinger Publications, Inc.
ISBN: 1879237342

Peaceful Dying

The step-by-step guide to preserving your dignity, your choice, and your inner peace at the end of life

By Daniel R. Tobin, M.D.
with Karen Lindsey

This practical guide to planning end of life care is based on the FairCare program for peaceful dying which Dr. Tobin developed at the V. A. Hospital in Albany, New York. He is now expanding the program to a network of other hospitals. The book takes a systematic approach to decision-making intended to increase autonomy and peace in end of life care. It is written in plain language for use by persons faced with terminal illness and their caregivers. It uses a 26-step program to help identify key decision points in choosing care. Major issues include development of advance directives, pain relief, choices in care settings, and compassionate advice on how to reach closure with loved ones. Overall it's a clear roadmap that is presented without excessive medical detail.

Perseus Books
ISBN: 0738200344

Who's Right (Whose Right?): Seeking answers and Dignity in the Debate Over the Right to Die

Edited by Robert C. Horn, III, PhD and Gretchen Keeler

The debate over one's right of choice, when it comes to end of life decisions, is certainly a hot one. No two people have the exact same opinion, yet thousands upon thousands of individuals and families face terminal decisions every day. This book offers readers the first truly objective look at all the issues from a unique perspective. The editors (one of whom is a 14-year survivor of ALS) provide interviews with 10 individuals who have faced terminal illness.

DC Press
Sanford, FL
(866) 602-1476
(Can also be ordered through www.alsa.org)
ISBN: 0970844425

Bereavement Books & Guides

A Grief Observed

By C.C. Lewis
A more religious account of rediscovered faith.
Bantam Books, New York
ISBN: 0583274864

A Guide to Understanding Guilt During Bereavement

By Robert Baugher
Caring People
ISBN: 0963597515

Caregiving and Loss: Family Needs, Professional Responses

Edited by Kenneth Doka and Joyce Davidson
With approximately 25 million family caregivers in this country, one out of four households are providing care for a loved one. It is important for healthcare professionals to understand the unique needs of family caregivers and offer compassionate support. Featuring writings from 13 nationally recognized experts in the field of caregiving and loss, this book is developed in conjunction with HFA's award-winning Living With Grief series.

Hospice Foundation of America
(800) 854-3402
ISBN: 1893349020

The Courage To Grieve

By Judy Tatelbaum
Harper and Row, New York
ISBN: 0060977859

Don't Take My Grief Away From Me

By Doug Manning
A warm conversational style takes the reader through all the emotions and experiences that accompany the death of a loved one. The first section of the book deals with those first few days after a death and all the plans and decisions that need to be made. The second section picks up the grief journey and provides guidance, assurances and hopes for healing.

ALS. Three letters that change people's lives. FOREVER.

Harper San Francisco
 (Also available in audiocassette format)
 ISBN: 0060654171

The Eyes Are Sunlight: A Journey Through Grief

By Shirley Koers
 Walker and Company
 ISBN: 0802725864

Grief Expressed When A Mate Dies

By Marta Felber
 Personal expressions, experiences and suggestions with space for one's own thoughts and feelings as well. "Workbook" format for journaling.
 LifeWords, West Fork, AK
 ISBN: 0965396746

The Grief Recovery Handbook

By John W. James and Russell Friedman
 Harper and Row
 ISBN: 0060915862

The Grieving Time

By Anne M. Brooks
 A spouse's personal account of the first year after her husband's death. Written in brief sections, and talks about loneliness, depth of grief, etc.
 Harmony Books, New York
 ISBN: 0517572222

Healing Your Grieving Heart: 100 Practical Ideas

By Alan D. Wolfeld, Ph.D.
 Companion Press, Fort Collins, CO
 ISBN: 1879651122

How It Feels When A Parent Dies

By Jill Krementz
 Alfred A. Knopf, New York
 ISBN: 0394758544

How To Go On Living When Someone You Love Dies

By Therese A. Rando, Ph.D.
 Bantam Books
 ISBN: 0553352695

The Journey Through Grief

By Alan D. Wolfeld, Ph.D.
 Companion Press, Fort Collins, CO
 ISBN: 1879651114

Life After Loss: A Personal Guide Dealing with Death, Divorce, Job Change and Relocation

By Bob Deits
 Fisher Books
 ISBN: 1555611893

Living With Grief When Illness is Prolonged

By Kenneth Doka
 Hospice Foundation of America
 Washington, DC
 ISBN: 1560327030

Living When a Loved One Has Died

By Earl A. Grollman
 Very poignant poems and brief thoughts, easy to read.
 Beacon Press, Boston
 ISBN: 0807027154

When Bad Things Happen to Good People

By Harold S. Kushner
 Inspired by the death of his 14-year-old son and his family shared ordeal, Rabbi Kushner tells how to deal spiritually with an unfair loss or tragedy.
 Avon Books
 ISBN: 038067033X

When Parents Die: A Guide for Adults

By Edward Myers
 Penguin
 ISBN: 0140092110

When Your Spouse Dies

By Cathleen L. Curry
 This book deals with a variety of practical concerns for those who have lost their mates to death, including stages of grief for adults and children, mourning, loneliness, sexuality, networks of support, financial priorities and planning, good health practices, and healing.
 Ave Maria Press
 ISBN: 0877934169

Understanding Grief: Helping Yourself Heal

By Alan D. Wolfeld, Ph.D.
 Readers are asked specific questions about their grief journeys and encouraged to think about and write down their responses.
 Companion Press, Fort Collins, CO
 ISBN: 1559590386

Personal ALS Stories

A Passion for Life

By Paul Brock

This new book is the story of Paul Brock who at the age of 53, happily married with three children, at the peak of an illustrious career, and a bundle of energy—except for a weak right forearm—was diagnosed out of the blue with motor neuron disease and given three or five years to live. Now 60, Paul writes, "I can no longer walk, play the piano, scratch my nose, cuddle my wife, hug my kids..." But he can still think, talk and propel himself by motorized wheelchair, love, be loved, laugh, cry, feel, work, dream, get cranky, empathize, hope, feel the touch of hand, taste the warmth of red wine, and many other things. Despite it all, says Paul, "I retain my passion for life. And for telling stories." This book is an incredible and remarkable story—a celebration of courage and tenacity of the human spirit.

ABC Books

Sydney, NSW, Australia

1-300-360-111

sueanna@opusnet.com.au

www.abcshop.com.au

ISBN: 0 7333 1447 3

Closing Comments—A Spiritual Journey into the Heart of a Fatal Affliction

By Brian A. Smith

This book focuses on the destination and sheds light on the path so that the cared-for and the care-giver are both illuminated on the way.

Clements Publishing

Toronto, ON

www.clementspublishing.com

ISBN: 1894667069

Cries of the Silent

By Ellen Bell

A touching and personal account of Evelyn Bell's life with ALS.

ALS Society of Alberta

Calgary, AB

www.alsab.ca

ISBN: 0968538304

Falcon's Cry

By Major Michael Donnelly, USAF, Retired

With Denise Donnelly

A moving memoir of the author's experiences as an air force pilot throughout the 1980s and the Persian Gulf War, that also confronts his postwar diagnosis of ALS. (888) 749-6342

(Can also be ordered through www.alsa.org)

ISBN: 0275964620

For Words—A Journal of Hope and Healing

By Chris Vais, BA, MDiv, DD

A spiritual experience of living with a terminal illness, ALS. Chris was a minister, but those of any faith or no faith background will benefit from "For Words."

www.als.ca/chris-vais/

ISBN: 0973347406

His Brother's Keeper: A Story from the Edge of Medicine

By Jonathan Weiner

Featuring ALS Therapy Development Foundation (ALS-TDF) founder James Heywood and his brother Stephen, this book tells the story of three epic lines of research that are coming together through the eyes of Jamie, a young entrepreneur, who gambles on a new model of therapy development to save his brother Stephen's life. The story also profiles the Heywood family as they defiantly fight ALS.

HarperCollins Publishers

ISBN: 006001007X

How Will They Know If I'm Dead?

By Robert C. Horn, III

GR/St. Lucie Press

2000 Corporate Blvd., N.W.

Boca Raton, FL 33431

ISBN: 1574440713

In Dreams—A Life Journey in Prose and Poetry

By Elizabeth Grandbois

Elizabeth Grandbois tells of her life story including her inspirational battle with ALS through prose and poetry.

Manor House Publishing, Inc.

(905) 648-2193

ISBN: 0973195606

Journeys With ALS

By David Feigenbaum

In this book you will find 33 journeys about people living with ALS. Some are hopeful, some sad. A few are angry. All are powerful, real-life examples of people doing their best to cope, often with humor and high spirits.

DLRC Press

P.O. Box 1061

Virginia Beach, VA 23451

(800) 776-0560

lynn@davidlawrence.com**Learning to Fall: The Blessings of an Imperfect Life**

By Philip E. Simmons, Ph.D.

Read an excerpt at www.learningtofall.com,Xlibris: www.xlibris.com, or by phone 888-795-4274 (during business hours, Eastern time).

In Learning to Fall, Lake Forest English Professor Philip Simmons tells the story of his spiritual journey, which began when he was diagnosed with the fatal Lou Gehrig's disease at age 35. With wisdom and humor, he finds answers to life's deepest questions and shows us how, against all odds, to live lives of depth, compassion and courage.

ISBN: 073884022

Letting Go: Morrie's Reflections on Living While Dying

By Morrie Schwartz

Doubleday & Company, Incorporated

ISBN: 0385318790

Making Sense Out of the Senseless-The McFeat Family ALS Journey

By Ruth L. McFeat

This book is the story of the McFeat family's 20 month journey with ALS. Ruth's husband Forrest had ALS and was cared for at home until his death. The purpose of the book was to express individual physical and emotional family members' journeys in an attempt to help others understand what it is like loving and caring for a person with ALS at home as well as provide practical information to those who are also facing a journey with ALS.

Proceeds from the sale of the book go to the Robarts Institute for ALS Research in memory of Forrest McFeat. Books can be ordered for \$10 plus \$5 for

postage and handling by writing the author at P.O. Box 124, Dutton, ON, N0L 1J0.

ISBN: 0968539408

My Luke and I

By Eleanor Gehrig and Joseph Durso

This is a moving story of an era and a love that can never be forgotten-by the widow of one of baseball's greatest heros.

Thomas Y. Crowell Company

ISBN: 0690011091

On Any Given Day

By Joe Martin and Ross Yockey

ISBN: 0895872331

On Eagle's Wings-Fulfilling the Needs of Your Terminally Ill Loved Ones

By Connie Bobo

This is an informative book giving helpful hints on ways of taking care of a loved one with ALS at home. Ms. Bobo's husband died of ALS in the prime of his life.

The ALS Association of Nevada

(702) 248-4507

ISBN: 0966541804

Tuesdays With Morrie

By Mitchell Albom

Story about Morrie Schwartz, professor at Brandeis University who died of ALS in 1995. Mitch was his student and visited him many times before his death and together they wrote this tender book.

Bantam Doubleday Dell Publishing Group, Inc.

1540 Broadway

New York, NY 10036

ISBN: 0385484518

When the Music Stopped I Kept on Dancing

By Angela Riggs

Written by a young woman about how she dealt with her ALS, managed her life and the art of living for the sake of life itself.

BookPartners, Inc.

P.O. Box 922

Wilsonville, OR 97070

(800) 895-7323

Fiction

Personal Injuries

By Scott Turow

This novel is about a successful personal injury attorney who is discovered by a federal prosecutor to be involved in bribing judges. The twist to this book is that the protagonist's wife is a person with ALS.

Farrar Straus & Giroux

ISBN: 0374281947

Books for and about Children

Living With Grief: Children, Adolescents, and Loss, (2000) edited by Kenneth J. Doka, features articles by leading educators and clinicians in the field of grief and bereavement. The chapters entitled "Voices" are the writings of children and adolescents. The book includes a comprehensive resource list of national organizations and a useful bibliography of age-appropriate literature for children and adolescents. Below is a list of current chapters and authors.

Bereaved Children and Teens-A Support Guide for Parents and Professionals

By Earl. A. Grollman

Bereaved Children and Teens is a complete resource for parents and professionals seeking to help children cope with the death of someone they know.

Beacon Press

25 Beacon St.

Boston, MA 02108-2892

ISBN: 0807023078

(800) 733-3000

Healing the Grieving Child's Heart: 100 Practical Ideas for Families, Friends, and Caregivers

By Alan Wolfelt, PhD

This book is for those in need of practical, day-to-day "how-tos" for helping grieving children they love. All ideas presented remind that grieving children need our unconditional love, support, and presence.

www.centerforloss.com

Healing Your Grieving Heart: 100 Practical Ideas for Kids

By Alan Wolfelt, PhD

This book is for young and middle readers (six-12 years of age) grieving the death of someone loved.

The text is simple and straightforward. There are many age-appropriate activities as well as gentle, healing guidance throughout.

www.centerforloss.com

In My Dreams...I Do!

By Linda Saran

Inspired by the author's mother who continued to nurture an intimate relationship with her grandchildren despite her battle with ALS. Share the bonding experience of reading about two sisters who escape the world of their grandmother's physical limitations to the freedom of her dream realm where anything is possible. Together, the girls explore a place where bodies are limited only by mind. This narrative gives testimony to the magical power of the imagination - a secret that most children instinctively possess...and many adults. It has universal appeal to all navigating the road of life together.

ISBN: 0-9672082-0-3

Available through www.lesturnerals.org for \$12.95 US

Lou Gehrig: The Luckiest Man

By David A. Adler

ISBN: 0152005234

Mr. Adler is the award-winning author of more than one hundred books for children. This beautifully illustrated book by Terry Widner depicts the life and legend of Lou Gehrig. For children of all ages

Gulliver Books

Harcourt Brace & Company

525B St., San Diego, CA 92101

15 E. 26th St., New York, NY 10010

(800) 544-6678

Grandpa, What is ALS?

ALS Society of Alberta

400, 320-23 Avenue S.W.

Calgary, AB T2S 0J2

(403) 228-7752

When Someone Has a Very Serious Illness: Children Can Learn to Cope with Loss and Change

by Marge Heegaard

Woodland Press

99 Woodland Circle

Minneapolis, MN 55424

(612) 926-2665

ISBN:0962050242

ALS. Three letters that change people's lives. FOREVER.

When Someone Very Special Dies: Children Can Learn to Cope with Grief

by Marge Heegaard
Woodland Press
99 Woodland Circle
Minneapolis, MN 55424
(612) 926-2665
ISBN:0962050202

Saying Goodbye: Bereavement Activity Book

By Jim & Joan Boulden
Boulden Publishing
P.O. Box 9249
Santa Rosa, CA 95405
(800) 238-8433
ISBN: 1878076124

Talking About Death: A Dialogue Between Parent and Child

By Earl A. Grollman
How do you explain the death of a loved one to a child? Earl Grollman's book provides sensitive and helpful advice for families coping with loss. A compassionate guide for adults and children to read together, featuring a read-along story, answers to questions children ask about death, and a comprehensive list of resources and organizations that can help.
Beacon Press, Boston
ISBN: 0807023736

How to Help Children Through a Parent's Serious Illness

By Kathleen McCue
St. Martin's Press, New York
ISBN: 0312113501

The Grieving Child: A Parent's Guide

By Helen Fitzgerald
Simon and Schuster, New York
ISBN: 0671767623

Straight Talk About Death for Teenagers

By Earl A. Grollman
If you are a teenager whose friend or relative has died, this book was written for you. Earl A. Grollman explains what to expect when you lose someone you love.
Beacon Press, Boston

ISBN: 0807025003

Lifetimes

By Bryan Mellonie and Ropbert Ingpen
Excellent book about the life cycle for younger children.
Bantam Books, New York
ISBN: 0553344021

VIDEOS

The ALS Association's Living with ALS Video Series:

- **Clinical Care Management Discussion with ALS Experts**
- **Mobility, Activities of Daily Living, and Home Adaptations**
- **Adapting to Breathing Changes and Non-Invasive Ventilation**
- **Adjusting to Swallowing Difficulties and**
- **Maintaining Good Nutrition**
- **Communication Solutions and Symptom Management**

(Available at no charge to persons living with ALS, family members, and their personal healthcare professionals through ALSA at www.alsa.org)

ALS Lou Gehrig's Disease

This video is a 30-minute program from "The Doctor is In" a Dartmouth-Hitchcock Medical Center Production. It studies the cases of two people with ALS and a family caregiver. Expert medical and scientific commentary is provided by Lucie Bruijn, PhD, Research Director and Vice President of The ALS Association, and Jeffery Cohen, MD, a neurologist at Dartmouth-Hitchcock Medical Center.
Also available on DVD.

(800) 257-5126

www.films.com

Bearing Witness-Robert Coley-Donohue

This 90-minute film is an in-depth, poignant, and personal look at five years in the life of Robert Coley-Donohue, a Vancouver Island resident who lost his

wife to ALS and then received the same diagnosis himself later. The film depicts everyday life for Robert chronicling his progression, the support of his family and friends, the complexity of care, and hope and healing as he prepares to die.

National Film Board of Canada

(800) 267-7710 (Canada)

(800) 542-2164 (US)

(514) 283-9450 (International)

e-mail: international@nfb.ca

www.nfb.ca

Making Hard Decisions

Two videos dedicated to the memory of Dr. Barry Smith who died with ALS in June 2001. One video is designed for caregivers, either in groups or individually. The second video is designed for health care professionals.

Both come with a STUDY GUIDE to assist group leaders in using the videos.

To order, visit www.als.ca.

Palliative Care: Facing the Fears, Making the Journey

This video can be used to heighten awareness about palliative care and address some of the common questions and concerns about it. It is a useful visual aid to help explain palliative care to patients and families, and for the general public. English and French versions are included on each tape.

www.chpca.net/publications/informational_video.htm

The Man Who Learned to Fall

Produced by Beitel/Lazar Productions Inc. (2004)
McGill University

This is a feature documentary by Garry Beitel about a gifted writer and eloquent teacher who celebrates the wonder of life even as he is slowly dying of ALS. As his muscles deteriorate and his body becomes increasingly paralyzed, Phil Simmons continues to "wrestle joy from heartbreak" again and again and again at each stage of his ongoing losses. This film is an intimate portrait of Phil and his family and the odyssey they shared over the final months of his life. Phil Simmons is the author of the book, *Learning to Fall*, listed in this Resource section.

For information on how to order, cost and shipping, contact Beitel.Lazar@videotron.ca, or (514) 487-9726.

Ventilation: The Decision Making Process

A 20-minute video designed for ALS patients, their family members and health professionals. It includes interviews with three ventilator dependent ALS patients, family members and the medical staff from the Lois Insolia ALS Center at Northwestern University Medical School. (Available through the Les Turner ALS Foundation, www.lesturnerals.org).

Hospice Foundation of America Videos:

(800) 554-3402

www.hospicefoundation.org

Caregiving and Loss: Family Needs, Professional Responses

2001 Teleconference

This conference explores the ways that professionals can better understand and respond to the needs of family caregivers. The conference will emphasize issues such as the contexts of caregiving, as well as governmental and work policies that influence the experience of caregiving. Particular attention will be given to creating strategies that professionals can use to assist families in responding to the challenges and opportunities of caregiving, inform families as they make critical decisions about end-of-life care, and support families as they cope with their grief.

Living With Grief: Children Adolescents and Loss

2000 Teleconference

While we often discuss how we grieve as adults, rarely do we consider the losses that children and adolescents must face. Whether they are grieving the death of a parent or grandparent, or facing the losses involved in re-locating or divorce, children and adolescents often do not know how to cope. This teleconference considers the range of child and adolescent loss, as well as ways in which developmental level may affect responses to loss. It will present practical advice and intervention techniques to use to empower children and adolescents with effective coping skills.

Living With Grief: At Work, At School, At Worship

1999 Teleconference

Whether at work, at school, at worship or at home, grief not only affects our moods and motivation but our ability to function and our relationships as well. This teleconference considers the ways that grief influences us in varied settings, offering humane and practi-

cal suggestions to workplaces, schools and places of worship as to how they can assist those struggling with illness and loss. The panel features: Kenneth J. Doka, PhD, Michael Jemmott, MDiv, Michael Kirby, MSW, and Marcia E. Lattanzi-Licht, RN, LPC.

Living With Grief: Who We Are, How We Grieve
1998 Teleconference

An exploration of how factors such as culture, spirituality, gender and age influence the ways we experience grief and express loss. Living With Grief: Who We Are, How We Grieve features discussions of how culture and spirituality can either help or hinder the grieving process, as well as how patterns of coping with grief vary by gender, class, age or developmental levels. The panel features: Ronald Barrett, PhD, Kenneth J. Doka, PhD, Bernice C. Harper, MSW, Patricia Murphy, RN, PhD, FAAN, and Bradley Stuart, MD.

Living With Grief: When Illness Is Prolonged
1997 Teleconference

What are the specific issues of grief and bereavement during and after chronic illnesses? Living With Grief: When Illness Is Prolonged explores the particular stresses posed by different diseases such as cancer, AIDS, and Alzheimers. In addition, it focuses on the common problems that griever may experience throughout and after the illness. The panel features: Betty Davies, PhD, Kenneth J. Doka, PhD, William Lamers, Jr., MD, and Therese A. Rando, PhD.

INTERNET RESOURCES

The ALS Society of Canada does not endorse and is not responsible for the content of external sites. If you find a good internet resource you think should be added to the list, contact the ALS Society to make your suggestion. The Society will review it and if deemed appropriate, it will be added to the next updated edition of the Manual.

ALS Specific

ALS Society Provincial Partner Sites in Canada:

- www.alsab.ca
ALS Society of Alberta
- www.alsbc.ca
ALS Society of British Columbia
- <http://www.alsmb.ca>

ALS. Three letters that change people's lives. FOREVER.

- ALS Society of Manitoba
- www.envision.ca/webs/alsnl
ALS Society of Newfoundland and Labrador
- www.alsont.ca
ALS Society of Ontario
- www.sla-quebec.ca
ALS Society of Quebec

Other ALS Organizations:

- www.alsmndalliance.org
International Alliance of ALS/MND Associations - This site includes a directory of ALS/MND organizations worldwide as well as research reports from the latest International Symposium on ALS/MND
- www.alsa.org
The ALS Association (US)
- www.als.net
The ALS Therapy Development Foundation (Based in Cambridge, MA)
- www.lesturnerals.org
The Les Turner ALS Foundation (Chicago-based ALS organization)
- www.march-of-faces.org
March of Faces - raising awareness project
- www.mdaua.org/disease/als.html
Muscular Dystrophy Association (MDA) (US) also supports people with ALS and funds ALS research
- www.mnda.org
Motor Neurone Disease Association (UK)

Resources for Children:

- www.march-of-faces.org/KIDS/moe1.html
March of Faces space for kids
- www.alsindependence.com

Primary Lateral Sclerosis - PLS:

- www.geocities.com/freyerse
- maintained by an individual with PLS

Basic Information - Sponsored by a Research Organization:

- www.nlm.nih.gov/medlineplus/amyotrophiclateralsclerosis.html
National Institutes for Health (US)
- www.ninds.nih.gov/health_and_medical/disorders/amyotrophiclateralsclerosis_doc.htm
National Institute for Neurological Disorders and Stroke (Part of NIH)
- www.rarediseases.org
National Organization for Rare Diseases (NORD) (US)

- www.bcm.edu/neurol/struct/als/als1.html
Baylor University (MND/ALS Center--Team Approach)
- www.neurologychannel.com/als/

ALS Research:

- www.wfnals.org
The World Federation of Neurology ALS Site
- <http://www.columbiaals.org/>
The Eleanor and Lou Gehrig MDA/ALS Research Centre
- <http://www.hopkinsmedicine.org/alscenter/index.cfm>
The Robert Packard Centre for ALS Research at Johns Hopkins
- www.als.net
The ALS Therapy Development Foundation
- www.umassmed.edu/outcomes/als/
The ALS C.A.R.E. Program is a voluntary, physician-directed program to improve clinical outcomes for patients diagnosed with ALS

ALS Clinical Trial Database

- <http://clinicaltrials.gov>
US government site--Mainly US-based trials
- www.als.net
ALS-TDF compiles information on current clinical trials world-wide. Click on "Patients"

About Rilutek - manufacturer site -

- <http://www.alsinfo.com/index.jsp>.

Individual's web sites:

- www.alsindependence.com
Maintained by George Goodwin,
Person with ALS (Canada)

Assistive Technology

- <http://www.tetrasociety.org/>
TETRA Society
A voluntary organization of Canadian and US engineers and technicians who help to solve your assistive devices challenges
- www.abledata.com
ABLEDATA
Information on assistive technology and rehabilitation equipment sponsored by the US National Institute on Disability and Rehabilitation Research

Caregiver Sites

- www.howtocare.com
How To Care

- www.chpca.net
Canadian Hospice and Palliative Care Association (CHPCA)
- www.allianz.ca
Allianz Insurance Company features information for caregivers as well as holds an annual caregiver recognition contest.
- www.ccc-ccan.ca
Canadian Caregiver Coalition
- www.fcns-caregiving.org
Family Caregivers Network Society
- www.caregiver.on.ca
Caregiver Network
A Canadian resource dedicated to making caregivers' lives easier.
- www.caregiving.com
Online support service for caregivers of an aging relative, friend, or neighbour.
- www.ec-online.net
ElderCare Online is a beacon for people caring for aging loved ones. Whether you are caring for a spouse, parent, relative or neighbor, this site provides an online community where supportive peers and professionals help you improve quality of life for yourself and your elder. Includes chats for caregivers.
- www.care-givers.com
Empowering Caregivers offers comprehensive emotional and spiritual support for all caregivers through experts, articles, newsletter, journal exercises, spotlights, healing circle, resources, inspiration, humor and much more. It is about choices, healing and opening to love.
- www.von.ca
Victorian Order of Nurses
A national health care organization and a federally registered charity that has been caring for the lives and well being of Canadians in their homes and local communities since 1897.
- www.thefamilycaregiver.com
The Family Caregiver Newsmagazine, a Canadian publication which includes a resource directory, can be downloaded for free. You can also submit your personal caregiving story for consideration in an upcoming issue.

General Health Resources

- http://chp-pcs.gc.ca/CHP/index_e.jsp
Canada Health Portal Access to Authoritative and Trusted Information and Services
- <http://secure.cihi.ca/cihiweb/splash.html>
Canadian Institute for Health Information

- <http://www.canadian-health-network.ca/>
The Canadian Health Network - Health Info for everybody
- <http://www.health.discovery.com/>
Discovery Channel Health Site
- <http://www.medbroadcast.com/>
Med Broadcast-provides information on a variety of health topics including ALS (Click on "A" and scroll down until you see Amyotrophic Lateral Sclerosis)

Government Resources

- www.canadabenefits.gc.ca
Canada Benefits-Connecting You to Government Benefits
This site offers Canadian citizens government-wide information about financial benefit programs for individuals. Of particular interest may be the pages for seniors, people with disabilities and veterans.
- www.canadian-health-network.ca
The Canadian Health Network (CHN) is a new and growing network, bringing together resources of leading Canadian health organizations and international health information providers. The resources identified here will help you take care of yourself and the people you care about -- with tips on how to improve your health and well-being.
- www.cra-arc.gc.ca/disability
Disability Tax Credit (DTC) The "disability amount" on your income tax return reduces the amount of income tax that a person with a disability, or their supporting person, might otherwise have to pay. Visit the Government of Canada Web site for more information about this and about medical expenses you can claim.
- www.hc-sc.gc.ca/seniors-aines
Health Canada's Division of Aging and Seniors web site, with information on federal programs, statistics on aging in Canada and more.
- www.pwd-online.ca
Persons with Disabilities On-Line
Persons with Disabilities Online's mission is to provide integrated access to information, programs and services for persons with disabilities, their families, their caregivers, service providers and all Canadians, through the use of internet technology.

For information that is specific to the Province or Territory in which you live, visit your provincial government's web site.

ALS SOCIETY FACT SHEETS

Sheets from the ALS Society of Canada, store them here for easy reference. All Fact Sheets can be printed from the web site www.als.ca.

Included in the pocket page:

- #4 *Caregiver Stress: 10 Signs of Caregiver Stress*
- #5 *Reducing Stress: Reducing Caregiver Stress, Finding Better Ways to Feel Better*
- #8 *ALS First Steps-First Steps for Families*
- #9 *Ways to Help: Assisting Families Living with ALS*
- #10 *ALS/MND-The Dangers of Unproven "Therapies"*
- #11 *Clinical Trials-A Primer*
- #12 *Natural Health Products - Making Sense of Complementary and Alternative Medicines*

ALS RESEARCH UPDATES

The *Research Updates* referred to in the *Manual* have been included in the pocket page with the *Fact Sheets*. As additional updates are published, request them from your provincial ALS Society Office, or download them from the web site, www.als.ca.

- #1- *Oxidative Stress Not the Whole Story*
- #2- *Rapid Diagnosis for ALS on the Horizon*
- #3- *ALS4 Gene Found*
- #4- *ALS Research Accelerates*
- #5- *Another Step for Stem Cells*
- #6- *Canadian Wins Milton Shy Award for ALS Essay*
- #7- *Misadventure in the Motor Neuron*
- #8- *Cognitive Deficits Provide Clues in ALS*
- #9- *Devoted Researcher Brings New ALS Lab to Canada*
- #10- *Help or Hindrance*

Please take a few moments to provide your feedback about the Manual. We would like to know how useful the Manual is to individuals and families living with ALS and how we can make it even better next time. If you have helpful hints for managing day-to-day living with ALS, please share them with us. We will include them in future publications.

Thank you for your time and contribution!

1. Please tell us how helpful you found each section of the Manual by placing a check mark in the appropriate box.

SECTION	Not at all helpful	A little helpful	Moderately helpful	Very helpful	Not Applicable
What is ALS?					
Coping with ALS					
Getting to Know the ALS					
ALS Disease Management					
Assistive Equipment					
Legal and Financial Issues					
Resources					

2. For you, what aspect(s) of the Manual are MOST helpful to you and why?

3. What are your suggestions for how to IMPROVE the Manual in future?

4. Do you have any corrections or additions you would like noted for future updates?

5. Are there any other comments you would like to add?

6. Are you a (check all that apply):

- Person with ALS Family member/friend Primary caregiver
- Healthcare professional ALS Society staff person ALS Society volunteer
- ALS Society donor Other _____

OPTIONAL:

Name _____

Address _____

City/Town _____ Postal Code _____

Phone _____ E-mail _____

Please return your evaluation by mail or fax to:

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 265 Yorkland Blvd., Suite 300
 Toronto, ON M2J 1S5
 (Fax: 416-497-1256)