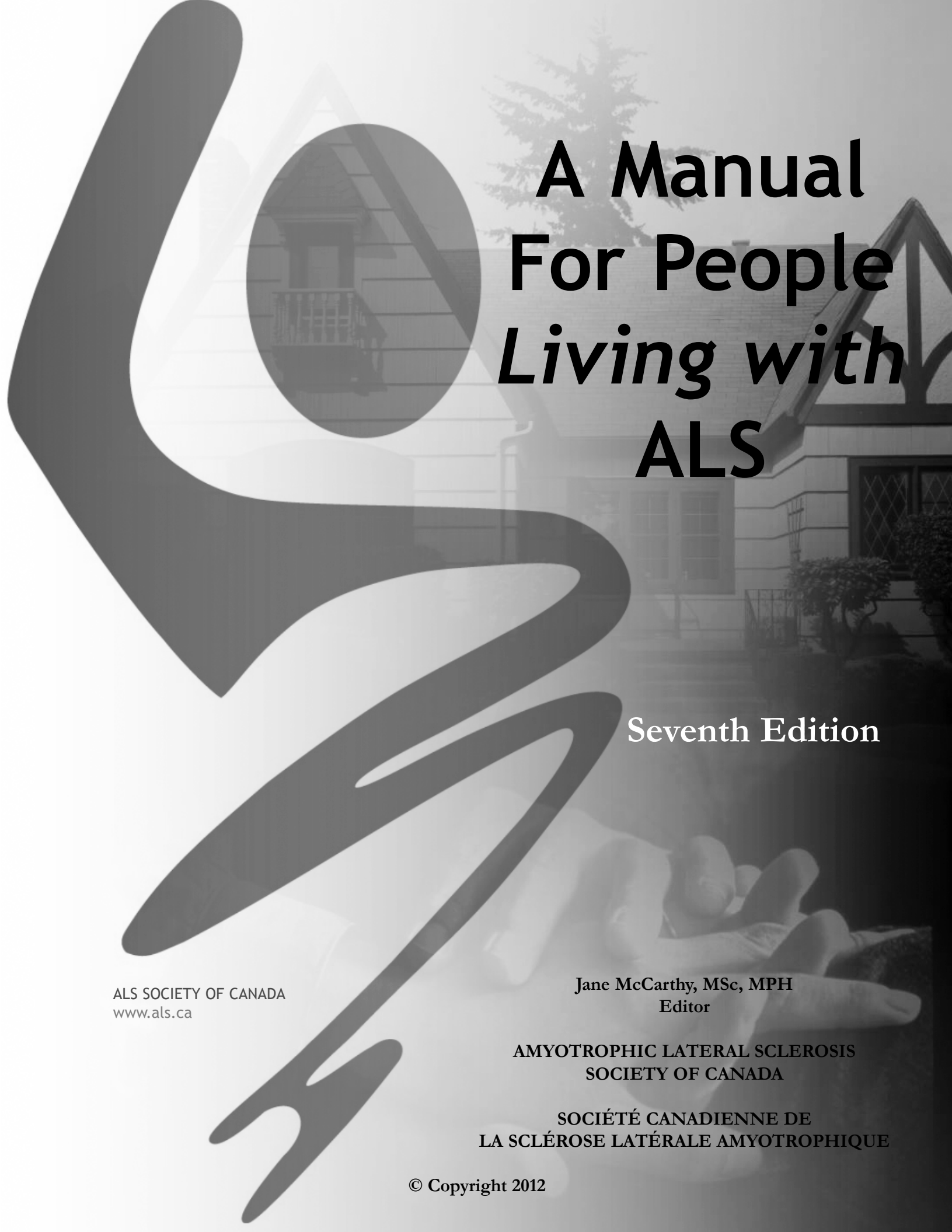

A Manual For People *Living with* ALS

Seventh Edition

Jane McCarthy, MSc, MPH
Editor

ALS SOCIETY OF CANADA
www.als.ca

AMYOTROPHIC LATERAL SCLEROSIS
SOCIETY OF CANADA

SOCIÉTÉ CANADIENNE DE
LA SCLÉROSE LATÉRALE AMYOTROPHIQUE

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ALS SLA

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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.
See "HOW TO USE THE MANUAL" section.

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.

Updated editions of the Manual will be published periodically, so readers with suggestions for updating or changing content, or altering the format to be as user-friendly as possible, are asked to share their insights by sending an e-mail to alscanada@als.ca.

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.

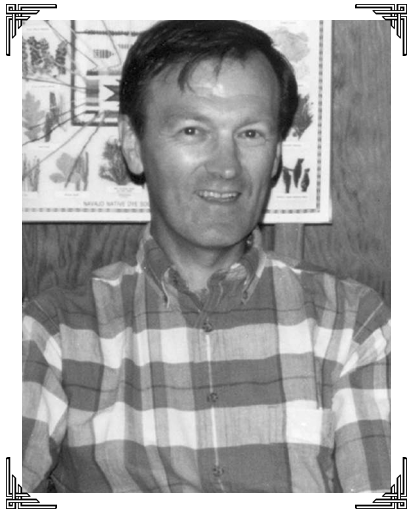
This was especially true for its original author Bob Macdonald.

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 still evident here in the seventh edition which reflects updated information thanks to the health care professional contributors who so graciously gave of their time to review the Manual and provide necessary changes (see "Contributors").

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.

-2012



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. Consider it to be a complement or additional resource to your primary source of information: your health-care team of professionals who have a special interest in helping you manage ALS.

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.

To get the most out of this resource, we suggest that you **think of it as your toolbox:**

Resource Tools

Review the "Resource Section" early to see what additional educational and support resources are available to help you learn more about ALS and specific aspects of coping with ALS.

Communication Tools

Use the Manual to share information about ALS with friends and family who are interested in knowing more.

Use information in the Manual to prompt discussions with your healthcare professionals about monitoring your progress and symptom management decisions.

Organization Tools

Use the copy-ready personal assistance tools included at the back of the "Coping with ALS" section to help you and your family caregivers keep track of your personal details, needs, communication records, healthcare professional contacts, equipment records, medications, and appointments.

Use the pocket page at the end of the Manual to store Fact Sheets and Research Updates you may download from the ALS Society of Canada web site and other pamphlets as you collect them.

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

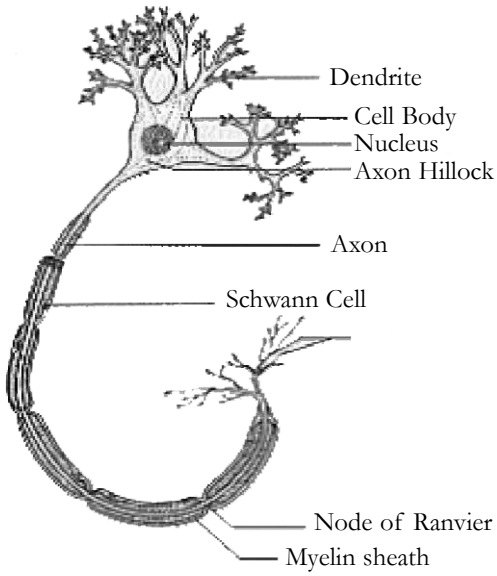
Dr. Jean-Martin Charcot, a French pathologist who founded the field of neurology, published the first full account of the symptoms of ALS in 1874. ALS is also known as Charcot's Disease, Lou Gehrig's Disease, and the most common form of Motor Neuron Disease (MND). In Canada, two to three people with 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 34 million, approximately 2,000 - 3,000 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. In people with the familial form of ALS, loss of function is usually more rapid in the legs, as opposed to in the arms, among those with sporadic ALS.

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, will likely 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.

NATIONAL NEUROMUSCULAR DISEASE REGISTRY

Epidemiological research, the study of disease trends within a population, helps to better understand a disease through the collection of information related to disease onset, progression, treatment and care, and demographic, environmental/lifestyle exposure factors.

Disease registries are database tools that provide an opportunity to collect this information to develop a risk profile for a disease or condition, how it develops, and how it responds to treatment and care.

In June 2011, a new national registry (the Canadian Neuromuscular Disease Registry (CNDR) for patients with neuromuscular disease was implemented. It will help patients connect with researchers to participate in clinical research that will benefit patients by offering possible new therapies, treatments and understanding of their disease. The CNDR is a Canada-wide database of patients who have been diagnosed with a neuromuscular disease. The term "neuromuscular disease" refers to a group of more than 40 diseases that affect how muscles and nerves work. ALS is the most prominent of these diseases in adults, and and Duchenne muscular dystrophy (DMD) is the most common pediatric muscular dystrophy.
The Canadian Neuromuscular Disease Registry (CNDR) includes 17 clinics across Canada located in Vancouver, Calgary, Edmonton, Ottawa, Toronto, London, Kingston, Montreal and Halifax.

Why participate in the registry if you have ALS? The Registry is the only means by which valid national epidemiologic data about ALS can be obtained. Patients with neuromuscular disease will benefit from this new national registry. The information that will be made available to researchers will lead to a significant increase in the number of studies leading to discovering the cause, treatment and cure of ALS. Finding treatments for neuromuscular diseases has been challenging, as patients are scattered across the country. This registry will allow doctors and researchers to look at medical data from large groups of patients helping them to find better ways to manage each disease.

All patients both adults and children across Canada who have been diagnosed with a neuromuscular disease are able to join the registry. Patients living outside the cities with affiliated clinics, or those not currently seeing a neuromuscular specialist, can register by contacting the CNDR National Office at the University of Calgary at 1-877-401-4494.
The CNDR is supported by the ALS Society of Canada, Jesse's Journey and the Marigold Foundation.

For more information about the registry please visit www.cndr.org.

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 and gene 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 several 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.

In addition to effects on the motor system in the brain, Sanjay Kalra, MD, from University of Alberta's faculty of medicine and dentistry released **two studies in 2011 that used advanced imaging to show that ALS attacks multiple parts of the brain and is not limited to the motor system.** Kalra used MRI scans to detect chemical changes that indicate different aspects of the type of degeneration seen in ALS, including death of neurons and scarring ("gliosis"). These advances have significant potential to track ALS and its progression, enabling development of more targeted treatments to slow or prevent the disease in

those parts of the brain which are affected beyond the motor system.

Environmental Factors

The new Registry mentioned above will allow for a potentially superior understanding of the role of environmental exposures as they relate to ALS. At present we know ALS is age-dependent, and is on the increase as the average age of the population increases. The role of an environmental factor is much discussed every time **clusters of ALS cases** have appeared to occur 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 Guam, the Auyu and Jakai of west New Guinea, and residents of the Kii Peninsula of southern Japan. 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). Additional results supporting a role for smoking in the development of ALS has been reported (Gallo et al., 2009).

A study of exposure to 11 classes of chemicals, or X-rays, provided evidence that there may be an increased risk for ALS with exposure to formaldehyde (Weisskopf et al., 2009).

There is a possible increased risk evident in those who have suffered a recent mechanical trauma. An association with electrical injury has even been postulated.

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 them in order to better understand the relationships and the new CNDR will help to do just that.

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.

Every newly identified and located ALS gene provides scientists a piece of the ALS puzzle and creates the opportunity to develop new animal models and cell lines that simulate these genetic abnormalities to identify 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.

In approximately 10% of people with ALS, the disease is inherited. There are now five different genes in which mutations are believed to be directly causative of ALS: SOD1, angiogenin (ANG), TARDBP, FUS, and FIG4. These genes together account for approximately one-third of all familial ALS cases.

Research to identify both categories of genes—directly causative or predisposing—represents a very dynamic field. A website is available which reports the most updated information:
http://alsod.iop.kcl.ac.uk/reports/reportSummary.aspx

In September 2011, a team led by Rosa Rademakers, PhD, from the Mayo Clinic Jacksonville identified the most common genetic cause known to date for ALS and frontotemporal dementia. Results show that a mutation of a single gene, called C9ORF72, accounts

for nearly 50 per cent of the directly inherited familial ALS and Frontotemporal Dementia (FTD) in the Finnish population, and more than a third of familial ALS in other groups of European ancestry. Further studies by other groups have found mutations in this gene in individuals with sporadic (i.e., non-hereditary) ALS. Identifying this defective gene provides important insights into the complex interplay between genetic risk for the disorder and other factors which contribute to disease onset and progression. These insights pave the way for a better understanding of ALS and FTD biology and the therapeutics that can be developed to counteract it. University of British Columbia's Ian MacKenzie, MD, was a key Canadian contributor to the study.

Genetic testing of the SOD1 gene, and the other 4 genes mentioned above, is commercially available. Testing for mutations in the SOD1 gene 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.

Testing at other known genetic loci is usually carried out for research purposes only. Even though some additional 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.

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.

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.

Presymptomatic testing for ALS-causative gene 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 or treatments 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. You may also 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 reactions, 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 detoxifying those that are normally produced.

In healthy individuals, special enzymes protect cells from dangerous free radicals. But **malformed enzymes, such as those found in ALS, may have the opposite effect, in essence initiating damage rather than protecting against attack by dangerous free radicals,** in a twisted game of molecular tag. Neil Cashman, MD, and colleagues at the University of British Columbia reported in 2011 on their use of a truncated enzyme and special antibodies to analyze the folding and misfolding of a key protein. **The goal is to create new proteins with binding capacity to act as a "sticky patch" where "bad" enzymes can attach and be removed from the system.** With further development, these proteins have the potential to block unhealthy interactions, thereby stopping disease progression in its tracks.

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 inflammatory response that could contribute to, and even perpetuate, a cascade of cell death in the nervous system.** The immune cells of the nervous system, called microglial cells and astrocytes, can respond to neural injury in a way that can be either beneficial or harmful. This microglial response which begins as a localized inflammatory reaction, has been implicated as a trigger of cell death pathways in motor neurons. While a mechanism that serves as a response to regional cell injury is useful on the small scale to clear away damage, it can become devastating on a large scale as secreted factors diffuse through the nervous system, killing adjacent 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 cell culture experiments and 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.

Neurotrophic factors were the first important category of molecules tested in large-scale clinical trials of ALS. These included brain derived neurotrophic factor (BDNF) and ciliary neurotrophic factor (CNTF). While these trials were not successful, it became clear that the question of delivering therapeutic reagents to motor neurons is a tremendous challenge which needs to be addressed. Recently, three other promising neurotrophic factors, glial-cell-line-derived neurotrophic factor (GDNF), vascular endothelial growth factor (VEGF), and insulin-like growth factor (IGF1), have shown promise in ALS mouse models. 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.

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 and other proteins in the motor neurons. NFs are believed to be responsible for maintaining the normal neuronal structure and shape. Studies making use of transgenic models to alter neurofilamentexpression reveal that abnormalities in the proportion 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. Peripherin, ubiquitin and the recently identified TDP-43 and FUS proteins are all being investigated for their roles in aggregate formation in ALS. Many other 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 examining this tissue in 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 to normalize the protein kinase ratios to prevent cell death from triggering.

By studying the spinal cords of people who died from ALS, Dr. Jean-Pierre Julien's team at Laval University in 2011 discovered an overproduction of a protein called TDP-43 in their nerve tissues. This protein can play a key role: when it is "overexpressed," it exaggerates
the inflammatory response that increases the vulnerability of nerve cells to toxic molecules that circulate in the body. The team is testing an inhibitor that could lead to the development of drugs to reduce this inflammation and partially restore the neuromuscular function.

Proteosomes and protein chaperones are cellular systems that shuttle proteins and digest them into their basic components in order to clear away damaged proteins in the cell. Researchers are investigating these systems 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 aggregate formation in motor neuron 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 glutamate binds at neuronal 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 somehow affect glutamate mechanisms.

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 technologically, 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 identifying 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: *Clinical Trials—A Primer*. This fact sheet can 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.

In 2008, ALS clinicians from 15 centres across Canada incorporated into a group called CALS (Canadian consortium for clinical research and trials). As a group,

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

CALS aims to bring clinical trials to ALS patients across Canada. Their first clinical trial to test the efficacy of lithium was carried out in partnership with a similar consortium from the eastern U.S. – NEALS. The study began in January 2009 and investigators were asked to halt the study following an interim data analysis in September 2009. While the results from the lithium clinical trial were disappointing in that the study was not able to duplicate the survival benefits seen in the original pilot study, the CALS centres continue to be sites of clinical trials for a number of promising ALS treatments. Currently, a stage 3 trial is in progress to study the efficacy of **ceftriaxone**, to see if it prolongs survival and/or slows decline in function in patients living with ALS. Ceftriaxone is an antibiotic in the class of cephalosporins that is approved to treat certain types of infections. Ceftriaxone may hold potential for ALS because the drug may also increase the level of a protein that decreases the level of glutamate - a potentially toxic nervous system messenger - near nerves, thereby protecting motor neurons from injury. Enrolment for this trial has been met. For more information on this trial: http://www.als.ca/en/research/clinical-trials/stage-3-ceftriaxone-clinical-trial-begins

A Phase III study to evaluate the efficacy, safety, and pharmacokintetics of **dexpramipexole** in patients with ALS is also underway in Canada. Six CALS centres are involved to study the effect of this compounds' neuro protective properties on slowing functional decline and extending survival. This multi-national trial is also being conducted in the US, Europe, and Australia. Enrolment for this trial has been met. For more information on this trial: http://www.als.ca/en/research/clinical-trials/phase-iii-clinical-trial-dexpramipexole-als

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 and to find out if you meet eligibility criteria to enrol in studies thatare recruiting subjects.

A database of worldwide clinical trial information can be found through the World Federation of Neurology at www.wfnals.org, or at www.clincaltrials.gov.

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**, subsection *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

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 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.

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

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 form 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 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.

Tips and Advice:

See the following Fact Sheets (available to download at www.als.ca/media/als_fact_sheet.aspx) and the *Resource Section* for helpful books and web sites.

First Steps for Families:
For families of those recently diagnosed with ALS

Ways to Help:
Assisting Families living With ALS

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 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.

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 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. **Resources for children available through the ALS Society include:**

See the following Fact Sheets:

Caregiver Stress: 10 Signs of Caregiver Stress

Reducing Stress: Reducing caregiver stress, finding ways to feel better

- When Someone Special has ALS-ABooklet for Children
- When your Parent Has ALS-ABooklet for Teens
- Helping Children Cope with ALS-AParental Information Guide
- 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 print resources.

The ALS Society of Canada has a section on its website called als411 which caters to youth with a parent with ALS. The first three resources listed above can be downloaded from the website, www.als411.ca. Additional booklets for school staff and health care professionals on talking with children can also be ordered in print or downloaded.

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.

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.

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 *Resource Section* 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.

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:

"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."

We communicate through several publications such as our *Manual for People Living with ALS*, research publications, brochures, fact sheets, booklets and our extensive web site: www.als.ca. 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 "Resources" section. Contact your provincial society office to order hard copies or download from the ALS Canada website.

In addition to being a source for 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, other useful resource sites, and much more. "Knowledge is power," so learn as much as you can!

Supporting Research

The ALS Society of Canada is **dedicated to finding the cause of and cure for ALS** through funding high-impact 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.

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 because of advances in neurology and other areas of science, we know that effective therapies and a cure are now, more than ever, within reach.

To learn about the kind of research the ALS Society is funding, see:
www.als.ca/research/our-research-program.

Providing Support

Support programs such as equipment loan programs, support groups, and other client support services are provided at the provincial level. Available **services vary by province.** Specific information on what is offered in your province may be included as an additional insert at the end of this Section. If you did not receive the *Manual* through a provincial society representative, please refer to the contact list below to find out what is available to you in your province and to register so you may benefit from what is offered.

At both the national and provincial levels there are **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, The WALK for ALS, 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 co-ordinated ALS Day at federal or provincial legislatures. Participation in Society activities can be very empowering.

Contacting the Society

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

ALS Society of Canada
393 University Avenue, Suite #1701
Toronto, Ontario
M5G 1E6
1-800-267-4257
www.als.ca

For programs and services available through the ALS Society in your province or territory, please use the following contact information:

Alberta and NWT
www.alsab.ca
(403) 278-2257
Toll Free: (888) 309-1111

British Columbia and Yukon
www.alsbc.ca
(604) 278-2257
Toll Free: (800) 708-3228

Manitoba and Nunavut
www.alsmb.ca
(204) 831-1510
Toll Free (866) 718-1642

New Brunswick
www.alsnb.ca
(506) 532-5786
Toll Free: (866) 722-7700

Newfoundland and Labrador
www.envision.ca/webs/alsnl

(709) 634-9499
Toll Free: (888) 364-9499

Nova Scotia
www.alsns.ca
(902) 454-3636
Toll Free: (866) 625-7257

Prince Edward Island
www.alspei.ca
(902) 439-1600
Toll Free: (866) 625-7257

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

Saskatchewan
http://alssask.ca

(306) 949-4100

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. Please ask the clinic coordinator at the ALS centre where you go for care to send your family doctor a copy of the CD-ROM, *Guide to ALS Care for Primary Care Physicians*. If you have chosen not to go to a specialty ALS clinic/centre for care, please contact your local Provincial ALS Society and ask them to send this resource to your doctor on your behalf.

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 in The "How To" Health Guide produced by the Health Charities Coalition of Canada at http://www.als.ca/en/publications-and-resources/care-resources/how-health-guide.

Primary Care / Family 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.

Palliative Care Doctor

A doctor with special training in pain management and other care aimed at improving comfort and quality of life for patients with incurable diseases is a palliative care specialist. One may be a member of your ALS team. If there is no palliative care specialist as a regular member of the clinic team you see at the ALS clinic/centre, you may eventually consult with one at some point in your disease. This type of doctor is also well trained in talking with patients about care decisions, including end-of-life issues.

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 Respirologist

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 conservation, 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 (diaghram) 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.

- **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 Health* sub-section.

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

- **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.

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

- **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 number 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.

- **Uncontrollable Emotions**

Uncontrollable emotions such as excessive laughing or crying may be experienced by some people with ALS. This is called emotional lability or pseudobulbar affect (PBA), and can be particularly frustrating. PBA is a condition that affects 15-45 per cent of people with ALS. PBA is characterized by sudden, involuntary outbursts of laughing or crying that do not match the individual's actual feelings. The reactions may be misunderstood by other people complicating matters. PBA usually catches people by surprise when it first happens; however, over time many people learn how to modify these emotional outbreaks and how to avoid some of the situations that trigger them. If you are experiencing this problem, talk to your doctor about medications that may help control it. The FDA recently approved a drug for PBA, called Nuedexta. It is a mixture of the cough suppressant dextromethorphan and quinidine, which is used to treat heart arrhythmias. It is expected to be available by prescription in early 2011. People with PBA may also 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 behaves 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 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 <u>your</u> 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 occu.pational 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. For more information refer to the ALS pamphlet on Sexuality, Intimacy and Chronic Illness. It can be found on the ALS website www.als.ca/publications-and-resources/als-fact-sheets.

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.**

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,

It is also recommended that you and your doctor read the **Fact Sheets -ALS/MND: The Dangers of Unproven Therapies**, written by Dr. Andrew Eisen and **Natural Health Products-Making Sense of Complementary and Alternative Medicines,** both listed in the Resource section of the Manual.

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.

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.

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.

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.
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)**

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

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.

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.

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.

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 education around safe handling and transfer techniques.

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.

It was previously believed that exercise was harmful for people with ALS due to damage from overuse. We now know that exercise can be very helpful. Exercise programs should be recommended by an occupational therapist or physical therapist to ensure the right kind of exercises and how much is right for you. Exercise should not cause pain and excessive fatigue or shortness of breath.

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

Four types of exercise may be considered for people with ALS.

1. Flexibility training - stretching and range of motion exercises (ROM)
- prevents contractures and frozen shoulder
- decreases spasticity
- stimulates circulation
- improves joint lubrication

2. Strengthening – moderate resistance exercises, for example, Thera-Band
- may decrease disuse atrophy
- may maintain mobility and function longer

3. Aerobic – exercises for example stationary cycling and walking)
- may slow deterioration
- reduces spasticity

4. Balance training
- may reduce falls

(Dal Bello-Haas et al., 2007; Drory et al., 2002; Krivickas, 2003)

The purpose of exercise for people with ALS is:

- To maintain or improve the strength and 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
- To help manage pain and stiffness due to spasticity and increase tone

Range-of-Motion (ROM) Exercises

A person with ALS needs to move each affected joint through a series of range-of-motion (ROM) exercises 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. This can be accomplish in different ways.

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 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. A physiotherapist can prescribe an exercise program that is right for you. They will demonstrate the exercises to 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. Use your fatigue levels to help guide you. Conserve your energy for activities you enjoy. Keep in mind that day-to-day activities, such as cleaning and cooking, count as exercise! 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.

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.

Develop an exercise program that fits into your daily routine. You may wish to break up your routine through the day or on alternate days to avoid fatigue. If you continue to experience fatigue, consult your therapist for a change in your program.

Participate in an active exercise program. It may be necessary to switch to active-assisted or passive exercises later. Your physiotherapist can help you to make decisions about the type of exercise that is most suitable for you.

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.

Exercises should not cause or increase pain. Consult your therapist if this happens.

MOBILITY AIDS

Mobility aids are used to:

- Prevent falls and related injuries
- Promote independent mobility

and contracture prevention when there is spacticity.

Injury Prevention

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

"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

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. If you have shoulder weakness, be careful that those assisting you do not pull on your arms, potentially causing injury to weakened joints. It is also 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 using a sheepskin, egg crate or tempra foam, gel mattress pad or an alternating pressure 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 sliding boards and mechanical 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. Walking poles have also started to become a popular choice. Gait aid prescription should be made in consultation with your doctor, occupational therapist, or physiotherapist. Mobility equipment will provide the stability you need and also help your walking be more efficient and less fatiguing. 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 forearm 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. Talk to your therapist about the right walker for you.

- **Orthoses**

Orthoses are devices that are attached to your body to support weak joints as well as aid in positioning

Orthoses help to maintain function and comfort. The most commonly recommended for people with ALS are ankle-foot orthoses (AFO), cervical (neck) collar; low-back brace; wrist splints, 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 hip muscle to lift the foot high enough to avoid tripping. However, this can cause fatigue in the leg muscles compensating. Use of an AFO can reduce the work of the hip and knee musculature thus enchancing endurance and reducing tripping and risk for falls.

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, or becoming too fatigued?

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. This which will require an assessment 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 if this is going to be required.

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, or for individuals with significant fatigue. Before purchasing a power chair you need to make sure your home is wheelchair accessible. Additionally, you may want to consider transportation of the wheelchair as most power chairs cannot fit in a regular vehicle. Most manual chairs can be folded into the trunk or back seat, making them a more portable option for most individuals.

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 appropriate overall seat width in order to go through doors at home.

A power wheelchair for someone with ALS usually includes: special drive control electronics which can be expanded as your needs change (e.g., if hands become too weak, hand controls can be changed to chin, head or foot, or single switch scanning; switches for operating seat tilt, recline, or elevation; adjustable back with head support, seat cushion; and power or manually operated elevated leg rests.

Not only can a wheelchair assist with your mobility, many everyday activities can be accomplished while seated in a wheelchair. This makes for fewer transfers

throughout the day, conserves energy and reduces the risk of injury. For example, trays can be affixed for meals or holding a laptop computer. Electronic switches used to operate the computer or any other environmental controls in your home can also be attached.

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 potential harm for the person being moved. The following transfer strategies can help, but it is important to always learn and practice transfers with a therapist before trying it on your own.

1. Transfers Without Equipment

Sit-to-stand transfer: When rising from a seated to standing position (whether alone or with help), proper positioning is imperative. Feet should be positioned flat on the floor, hip width apart, heels in line with edge of the seat. Scoot to the edge of the chair, lean your trunk forward 30-40 degrees and rise. Arm rests on the chair are a helpful additional support. There are a variety of handling techniques that can be shown to you and your caregiver if you require assist to stand. Some caregivers prefer the aid of a transfer belt. Your Physical Therapist can assist you and your caregiver in determining and teaching the most appropriate handling techniques. It is very important helpers do not grab and/or pull on your arms, especially if shoulders are weak, as this could damaged weakened joints. It is also important not to pull on the helper's back.

> **"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."**
>
> **Terry Schultz**
> **ALS family caregiver**

Standing pivot: This transfer is best suited to someone who is able to stand and take weight through the legs but may need some assistance achieving an maintaining a standing position.
Using either a transfer belt around the person's hips or placing your hands on their lower pelvis, bring the person's trunk forward and assist them into a standing position. Keeping your body close to the other person, assist them to step or pivot their feet towards the surface to which they are transferring.

Low Pivot transfer: This type of transfer is most useful for someone who can take some weight through the legs but not enough to fully stand or walk. When transferring to and from a wheelchair ensure that the brakes are locked and that the armrest has been removed. Place one arm around the person's back, just below the shoulder blades and the other arm under their thighs. Have the person lean forward and (if able) reach towards the surface to which they are transferring. Counting aloud throughout the transfer helps coordinate and minimizes the effort involved for both parties.

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 be taught how to assist you. Once you are on your side, bend your knees so that your legs hang over the edge of the bed and push yourself up into sitting through using the hand of your uppermost arm and the forearm of the other arm. A caregiver can offer stability as needed. To lie down from a sitting position the technique is reversed. Again, it is very important that caregivers do not grab and/or pull on your arms.

Repositioning in bed: When preparing to get into bed, always try to position yourself in such a way that when you lie down your head will be at the uppermost part of the bed. A Physiotherapist can provide teaching around bed mobility as needed. Bridging and rolling techniques can be useful for repositioning purposes. If you are unable to reposition yourself independently in bed a turning sheet can be a helpful tool for caregivers.

2. Transfers with Equipment

Transfer board: A smooth board acts as a bridge between two surfaces and is very helpful when

standing transfers are becoming difficult. With one end of the board under your buttocks, and the other placed onto the surface to which you are transferring, a sliding transfer can be performed with or without caregiver assistance. This technique is best used between surfaces of similar heights.

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.

Talk to Your Therapist

Your therapist can also provide information on other transfer devices such as the standing transfer pivot. Ask about the latest in available equipment and work with your therapist to determine what will best meet your needs. Remember, before attempting any transfers make sure your therapist has explained them and trained you and your caregivers.

- **Lifts**

Lifts are valuable assistive devices especially when you are no longer able to stand. Additional information on lifts is included in the Manual under the "Assistive Equipment" section.

TRAVEL TIPS

- **Facilities**

If you have problems walking, or are in a wheelchair, telephone your destination ahead of time to find out what accessible facilities are available where you will be staying, as well as at any of 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 a discounted rate. 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. **In particular if you need to you use a non-invasive ventilator while traveling, be sure to contact the airline before booking a trip to find out if they permit the device you use to be used in-flight.** You may need to contact the device manufacturer to see if they have a sticker you can affix to the device indicating it meets aircraft safety requirements.

- **Auto Travel**

Some families with ALS purchase a wheelchair accessible (barrier-free) van, for every day use as well as road trip vacations. Keep in mind you must be assessed and trained in the use of hand controls by a driver rehabilitation specialist, before you are legally able to drive a hand controlled vehicle. **There are companies that also rent barrier-free converted vans** for extended road trips, or to use upon arrival at your destination. 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.

- **Public Transit**

Many larger cities offer accessible public transit for people with mobility impairments. Accessible public transit typically consists of specially equipped buses or taxis licensed by the municipality to carry disabled passengers. Note that buses and taxis may require booking appointments for pick-up. Larger cities may also have subways accessible by elevator. Check with your provincial society for public transit options in your area.

- **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 need for adaptive aids will change overtime 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.

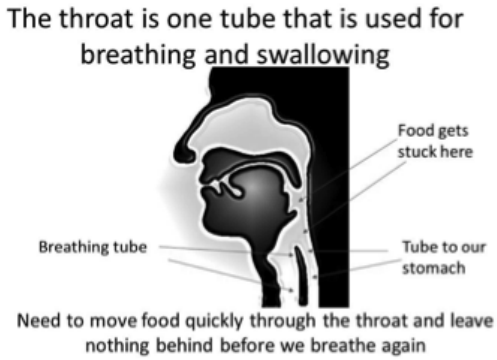
Fatigue plays a very big role in why a person's nutritional status declines. Weight loss occurs because with ALS, increased energy is required for all activities including eating and your body's basic energy requirements are higher (increased basal metabolic rate).

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 mouth, through the throat (the pharynx) and down the **esophagus** to the stomach. 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.

Weakness, slowness, and poor coordination in the muscles of the throat may cause material to stick in the throat or become misdirected into the voice box (i.e. the larynx). Aspiration is when food goes down the wrong way when we swallow. Our throat is tube with two openings on the bottom: one opening goes down to our stomach (the esophagus) and the opening goes to our lungs (the trachea). Aspiration typically happens because we have to hold our breaths when we swallow. If things get stuck in our throat when we swallow, it often gets sucked down into the lungs when we start breathing again. As food, drinks and saliva get stuck in the throat and sucked into the lungs, it can cause a lung infection (pneumonia)



The Four Phases of Swallowing

In **phase-one**, the food is chewed or "processed" in the mouth to form a **mass (bolus)**. 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. It can also lead to residue in the mouth (material left in the mouth after the swallow is completed).

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 or breathing tube).**

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 textures of

are easiest and safest 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.

Swallowing assessments sometimes include an x-ray procedure called a **Modified Barium Swallow Videofluoroscopic Swakkiwubg Study (VFSS)**. This is ordered with the permission of your doctor and is an outpatient exam. This is done because the speech-language pathologist cannot see what is happening inside your throat when you swallow. 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, testing if different positions or strategies help you swallow more safely and for determining what kinds of foods are safest for you to swallow.

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 during or after eating and drinking
- A wet or gurgled sounding voice during or after eating or drinking
- 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 mixing 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 abilities. 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. Smaller and more frequent meals may be easier than three large meals and lead to less fatigue.

- **Be relaxed when eating**

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 and sips**

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.

Sometimes saliva or mucus stuck in the throat, strong smells, smoke, alcohol, cold bursts of air, spicy foods or poorly control acid reflux (frequent indigestion) can cause the voice box to have a muscle spasm (laryngospam) which may temporarily block of the airway. Avoiding things known to trigger laryngospasms may be help. The use of medications to reduce muscle spasticity or tension and medications to prevent acid reflux or indigestion may eliminate or reduces the severity of the laryngospasm. Other techniques you can try if you experience a laryngospasm include sniffing through your nose, moving your chin upward and outward or a doing a variety of breathing technique to clear the throat of any residue.

- **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 esophageal 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

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:

- **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, speech-language therapist, or doctor regarding changes to your diet. With changes in your oral (mouth) skills (, 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. (e.g., pasta and casseroles).

- **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.(e.g. water, milk, juice). Water served at room temperature is often harder to swallow that ice cold water. For some people, carbonated water is swallowed more safely than non-carbonated water.

- **Avoid very dry foods** (i.e. dried meat, popcorn, nuts, hard candies may pose a choking hazard. 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** (e.g. watery soups mixed with semi-solid meat and or vegetables or cold cereals with a firm texture mixed with milk).

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 or natural thickeners
- Semi-solid foods (e.g., pudding, custard, cottage cheese, puréed fruit)
- Oral nutritional supplements (e.g., Ensure+ or Boost+)

See the "Resources" section for cookbooks with easy swallowing recipes. To view on-line the Meals for Easy Swallowing book produced by the MDA ALS Division, go to http://www.als-mda.org/publications/meals/

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. Constipation may be a problem – the best solution is to get enough fluids over the day. Refer to the ALS Fact Sheet "Constipation" at www.als.ca/publications-and-resources/als-fact-sheets.

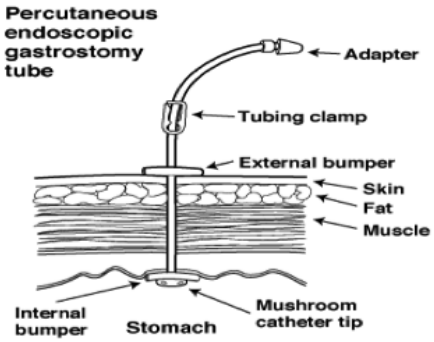
Many persons with ALS reach a point when it is difficult to meet nutrition needs by eating alone**.If you haven't already done so, now is the time to discuss with your doctor the option of having a feeding tube put in place to make sure you maintain good nutrition.**

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.



http://my.clevelandclinic.org/services/percutaneous_endoscopic_gastrostomy_peg/hic_percutaneous_endoscopic_gastrostomy_peg.aspx

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.**

SWALLOWING PROBLEMS: FOODS TO LOSE...	SWALLOWING SOLUTIONS: FOODS TO CHOOSE...
• 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)	• 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

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. To watch a video about PEG tube insertion and feeding produced by Sunnybrook Media, go to http://www.youtube.com/watch?v=yBzy3tHOps0&feature=related

- **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, and medication

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.
Tube feedings are typically started about 24 hours after the procedure and often the external bumper will need to be loosened before the first feeding. 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 practice 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, or by using a syringe. 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, reflux, or aspiration due to not sitting up enough, high feeding rate, 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 at 90 degrees during the feeding and for another 45 minutes after the feed is completed.

- **Ask for home care tips**

- **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. You will have to do water "flushes" as prescribed by your dietician before and after each time medications & PEG feedings are given. 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 (formula). Please note that the cost for these supplies is not usually covered. In addition, you might have to hire and pay for private care to administer your feeds if you or your family members are unable to do so. It is important to inquire ahead of time what will be supported in your community and what you and your family will be responsible for. If you cannot be supported at home, the option of going to a Long Term Care (LTC) facility might need to be considered.

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 or your dietitian 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

- **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.

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®, Nutren 2.0®, and Nutren® with Fibre, Isosource®, Resource 2.0®, and Compleat®. Your dietitian will prescribe what is best for you.

- **Making your decision about a feeding tube**

- **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 decision 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**, 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. Speak with your ALS Social Worker about appropriate paperwork which will need to be filled out according to the legislation in your province.

- **Use of feeding tube at end of life**

At end of life, either at home or in hospice, your palliative care team will likely discuss with you and your family the appropriateness and timing of tapering your intake of formula via the feeding tube. Towards end of life, the appetite and the body functions slow down and the need for nutrition decreases, hence, the volume of formula you are taking may be slowly decreased to ensure your comfort.

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 you use your natural speech functionally 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.

Please note that exercises to strengthen your oral muscles are contraindicated in ALS. These exercises may exhaust already weakened muscles, providing no benefit. Speech therapy for people with ALS does not focus on strengthening oral muscles, as the exercises may be tiring and hasten deterioration. Please consult an SLP in an ALS clinic to further discuss this issue as needed. 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 fasiculations (twitching) in your mouth and throat muscles
- Changes in voice quality (e.g. hoarseness, strained-strangled, lower pitch, nasality, breathiness, monotone, volume changes)
- Changes in your speech (e.g. nasal sounds, slower speech, slurred speech)
- Difficulty making your speech understood
- Increased difficulty with speech as the day goes on due to 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 able to naturally adjust to these changes and use communication strategies, but it might be more challenging for others to do so.

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 your 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

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 nasal regurgitation (food or liquids coming up into your nose). 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. It is also a custome made device and may not be covered by your insurance. You should consult a speech language pathologist and/or prosthodontist (dentist who specializes in oral devices) to determine your best option.

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
- Availability of support from family/friends
- Cost/coverage
- Portability
- Versatility
- Speed of communication
- Circumstances of use
- Amount of follow-up needed

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 needs with your immediate family and other caregivers
- Use non-verbal signals (e.g., eye blinks for yes/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 functional 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. Voice banking (recording your voice) can be considered before any speech changes occur, or when speech changes are still mild. This could allow you to use your natural speech, in combination with synthesized speech on a communication device at a later time should you need to use an AAC device

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 and/or a speech-language pathologist can help train you on 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

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 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. In addition, your communication partner can assist you by scanning through the rows and columns of an alphabet board to allow you to spell out words and sentences. You can select the letters by using a Yes/No system according to your abilities (e.g. eye-blinks, looking down, eyebrows movements, etc.)

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.

- **Other Signal Systems**

Low tech systems are available where you hit a switch or the device and it makes a sound (for example, a bell, intercom, or buzzer) 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 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.

In addition, if you leave home without a caregiver, it may be wise to wear a Medic Alert (medical emergency) bracelet.

- **Manual Communication Aids/Adapted Access**

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, laser 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 Devices with Recorded Messages**

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, hitting a botton or a switch, and you now have more messages available.

- **Specialized Telephone Equipment**

If you are unable to lift a phone receiver and/or dial a phone number, special phone services are available plus an **operator dialing service.** There is also a **hands-free speaker phone, activated by a voice commands, or by hitting a single switch.** 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 or AAC Team. These systems can be very expensive and there is a wide range of 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,

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 completion, word prediction, abbreviation expansion, and a menu of the most commonly used 100 or so words.

- **Word completion:** special software loaded in the system predicts the word you are starting to input after one or two letter selections. Typically three to eight possible words are displayed for selection. The options change each time a new letter is entered.

- **Word prediction:** upon completing a word, the special software predicts the next word based on commonly used phrases. Again, three to eight possible words are displayed for selection. Some systems even learn the words you use 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.
- **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 may be able to 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 other parts of the body using a switch, mouse, or joystick, head movement, or eyegaze. Your AAC team will be able to help you determine what access method is best for you.

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 (Speech Generating Devices (SGDS)**

Some of 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). Others now include email and internet options. They can be used on the telephone, with a group of people, or in quiet face to face conversations. Recently, portable devices such as the iproducts with speech output applications are also available. Although not specifically made for people with communication impairments, they offer a more affordable option if appropriate. These systems work well for individuals who still have good hand dexterity and strength.

- **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.

Computers are less portable than a device designed specifically for communication. Even laptops tend to be less portable because they are bulkier and more difficult to mount on a wheelchair than a dedicated communication device. However, iPads and e-readers are very portable. Computers can be slow to start-up and batteries do not last as long as the dedicated devices, but the iproduct batteries do.

- **Intergrated Systems**

Integrated systems combine a fully operational computer and face to face communication system all in one device. They allow multiple options for use that can adapt to changing needs, some of which include touch screen, standard hand held mouse, head mouse, switches and even a camera that tracks eye movement.

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.

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, coughing and swallowing safely 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 and coughing ability.

Poor breathing function leaves a person with ALS feeling fatigued, short of breath, and more prone to respiratory infections and respiratory failure. **Therefore, a vital part of the ALS disease management plan is to monitor breathing function throughout the course of the disease.** However, the reality is that the best way and when to address respiratory issues in ALS remains uncertain 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. Another publication you may find helpful, which includes anatomical diagrams and pictures of various ventilatory support equipment, is Breathe Easy: Respiratory Care in Neuromuscular Disorders. This is available through the MDA in the United States or as a download at http://mda.org/publications/PDFs/BreatheEasyBooklet.pdf.

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 **carbon dioxide** (CO2) is cleared from 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 exertion 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** and it is important to tell your doctor.

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 smaller breaths (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. You will need to consult with a skilled professional to train you and your family.

Quick List of Signs and Symptoms of Breathing Problems

- Breathlessness
- 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 is 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 breathing support like mechanical 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 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 160 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 generated 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 (SaO_2)

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 **(SaO_2)** 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 CO_2 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 (O_2/CO_2), 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.

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.

STAYING HEALTHY: PREVENTIVE AIRWAY MANAGEMENT STRATEGIES

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**. 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 pres sure 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-inva-**

Mechanical Ventilation: Things to Consider		
Type of Ventilation	**Non-Invasive Ventilation**	**Invasive Ventilation (with Tracheostomy)**
Advantages	• 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	• 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	• 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 or impossible to use • Gastric distention may occur • Most centres are familiar with traditional invasive ventilation however, few have experience with day time mouth piece ventilation	• 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 • Most people will be unable to speak or swallow • 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. A small number of patients who retain their ability to speak clearly, swallow easily and clear their airway effectively may be able to use a chair-mounted volume ventilator to assist breathing through a mouth-piece rather than a mask. This requires a respiratory team with experience in mouth-piece ventilation but may prevent the need for a tracheostomy and significantly prolong survival. If an individual becomes highly dependent on a ventilator and not able to breathe independently, then non-invasive ventilation will require a ventilator (or bilevel device) with alarms and an internal battery in case of power failure or disconnect.

Invasive Ventilation

Invasive ventilation requires a **tracheostomy**, an invasive surgical procedure. It is recognized that fewer than 5% of ALS patients choose to under-go tracheostomy placement. 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 ventilation 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 but most patients with ALS and a tracheostomy are unable to communicate verbally.

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 (see "Financial and Legal Considerations section) 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. This could put you and your family in a serious position which you may have chosen to avoid. One of the most difficult circumstances occurs when an individual with ALS fails to engage in this decision-making process and experiences invasive ventilation and ICU admission when, if properly engaged and informed, they would not have chosen to do so. We readily make

they would not have chosen to do so. We readily make plans for our finances in our wills; why should we not make clear decisions about our care while we are able? If at any time you decide you are interested in any form of breathing support, you should be referred to a respirologist.

Withdrawal of Invasive Ventilation

A very small number of individuals will accept invasive ventilation under any circumstances and others, rarely, on a temporary basis only if it is highly likely that they will recover and no longer need it. Invasive ventilation can and should be withdrawn if it is in any way against the wishes of the individual or if they have become "locked-in" and are unable to manage any communication. Any uncomfortable symptoms or pain or breathlessness will be managed with pain medications (narcotics) and sedatives (benzodiazepines). Managing symptoms, including those experienced in the withdrawal of ventilatory support are fundamental to the provision of Palliative Care.

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 Sensonic, 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.prodhelp.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 including organizing financial and legal affairs and determining your wishes for medical interventions 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 influenced 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. 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 such as 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 on your behalf without knowing your wishes. This may strain** relationships, cause unnecessary financial hardship, or result in a less peaceful, comfortable death.

An excellent resource to help you sort out your thoughts and engage conversation with loved ones is a card game activity called "Go Wish." It gives you an easy, even entertaining way to talk about what is most important to you. The cards help you find words to talk about what is important if you were to be living a life that may be shortened by serious illness. Playing the game with your relatives or best friends can help you learn how you can best comfort one another when you most need it. The game can be played on-line or you can purchase the deck of cards. To learn more go to http://gowish.org/index.php.

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 life sustaining 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**

Typically, over time, accepting that life is coming to a physical end may become less difficult. The functional and role changes associated with ALS often provide opportunity for reflection of one's past, present, and future. During reflection of one's life journey, feelings of greif and loss may be 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 sometimes, work our way back to some form of acceptance.

Instead of looking at oneself as dying, it may be easier to consider yourself as being in the final stage of life. Therefore, the focus 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**

For individuals are facing death, and those around them, uncertainty about what happens during the last days and hours of the dying process may cause fear and anxiety. To minimize these fears, and/or anxieties, 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 this way. With gaining acceptance and appreciation 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 to provide 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 an advance care plan 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 care 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 care 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 care plan to reflect your current outlook allows you, your alternate decision maker (Proxy, Representative, Power of Attorney for Health Care), and the healthcare 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 care 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 an **advance care plan** should help to ensure a dying person is treated according to their wishes and will have a dignified death as defined by them. Completing an advance care plan 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 an advance care plan 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*. An excellent on-line Canadian resource which describes advance care planning and making a plan is the Speak Easy Program at http://www.advancecareplanning.ca/home.aspx.

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

1. **Gather** information.
2. **Talk** about decisions.
3. **Prepare** an advance care plan.
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 an advance care plan are:

Resuscitation: Ask yourself whether or not you want to be resuscitated in the event of respiratory failure. Ask your doctor about how to obtain 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 care plan 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. Discuss with your health care team how you would like any pain controlled.

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 alternate decision making and their contact information, in the event of a med ical emergency outside the home.
5. Revisit your decisions every few months and re-evaluate. Communicate any changes you want to make.

In discussing the content of the advance care plan with your healthcare team, it is vital you explore the medical management of care of symptoms when the conventional intervention is not desired (e.g. invasive ventilation). In particular, when you choose not to have invasive breathing support, it is advisable to have a plan for managing feelings of breathlessness, and for the rare instance of acute breathing distress. Faced with the dilemma of acute breathing distress and with a plan indicating no life sustaining interventions, it may be reasonable to accept temporary non-invasive breathing support (e.g., bi-level ventilator) to control symptoms. Both invasive and noninvasive 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 life-sustaining therapies including mechanical ventilation and a feeding tube withdrawn. However, you may wish to speak 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 and healthcare team 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 Clinic or 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. 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: AGuide 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.

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 wells 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, AGuide to Recalling and Telling Your Life Story, published and available through the Hospice Foundation of America (www.hospicefoundation.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. 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 function. 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 your healthcare professionals, 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, **thick handle, lightweight untensils can make eating much easier for those with a weak grasp.** 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 slightly raised edge to a plate so that food can be pushed against it onto a spoon or fork. Common devices such as electric toothbrushes and shavers can make these tasks easier when arms are weak.

The double action of gripping and turning a doorknob may also be a problem for you. Changing to a **lever style handle** could be the answer since it allows the door to be opened by pushing down or pulling up the lever. A broad 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, Pen/pencil grips can be found at school supply stores. Gel pens that glide easier on paper may also help. Your health professional can show you different writing aids that minimize the amount of pinch or grip required to hold 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, such as zipper pulls and button hooks. **Velcro** is a popular replacement for buttons, zippersm and laces. Elastic shoe laces can turn lace up shoes into slip-ons. 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. Ensure pullovers are loose and stretchy. Your occupational therapist can show you tips to make dressing easier.

Electronic devices known as **"environmental aids to daily living (EADLs),"** can be installed in the home to enable a person with ALS to 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. Hands-free speaker phones can be used with voice commands or the touch of a switch.

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 useful 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, enhance comfort, and aid in function. Orthoses are typically prescribed by a **physiatrist**, and may be recommended by your doctor or physiotherapist or occupational therapist..

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).**

- **Hand and Wrist Supports**

A wrist and thumb splint can stabilize the wrist and thumb, helping you to grasp objects. Additional supports can be added for positioning the fingers to enchance your function despite weakened fingers.

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 "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 therapist which type of aid might work best for you. Another type of cane, or forearm crutch extends up the lower arm, almost to the elbow, with a ring that fits around the arm for added support. Walkers provide maximum support and stability because they spread the weight over a wide area. **The choice of cane or walker should be made in consultation with your doctor, occupational therapist, or physiotherapist. You should not try to use it until you have received instruction from your therapist.**

Wheelchairs

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.

- **Transport Wheelchairs**

Transport wheelchairs are helpful for people who require assistance or cannot walk long distances. They are very basic, provide minimal postural support, and require assistance to push. However, they are inexpensive, very portable, can be easily transported in a car or airplane.

- **Manual Wheelchairs**

Lightweight manual wheelchairs are often used by people who retain **trunk stability and the ability to self-position** themselves in the chair. Supportive seat and backrests can be added for comfort and support. They have large wheels that can be propelled by the user and push handles at the back if assistance is needed. These chairs are light to push and may be transported by car. Because ALS is a degenerative disease, it may be more cost-effective to rent or borrow this type of chair from your provincial ALS Society equipment pool, leaving funds available for other equipment.

- **Manual Positioning Wheelchairs**

Manual wheelchairs with dynamic positioning features, for example that tilt back, can be used for people with limited ability to weight shift or reposition. In most cases assistance is required to move and tilt the wheelchair. Specialized seating can be added to optimize postural support, comfort and pressure relief.

- **Power Scooters**

Power scooters can be used by people who have limited walking but can transfer independently and have adequate arm strength to operate the controls. They provide basic seating and postural support and are generally suitable for community mobility only. Therefore cost and length of use are important considerations.

- **Power Wheelchairs**

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.

Power wheelchairs allow a person to remain mobile and independent throughout the more advanced stages of ALS in their home and community. However, not all power wheelchairs are created equally! It is essential that modification and customization in seat and back supports, positioning features, electronics, and controls are possible for complex and changing needs.
Specialized seat cushions and back supports can be

added to optimize postural support, comfort and pressure relief. Power dynamic positioning features like tilt, allow independent repositioning for weight shift and pressure relief to prevent pressure sores and postural changes to optimize respiration and comfort. Some power wheelchairs have capabilities to control other devices like your TV or computer, for example. If hand and arm function are limited and you can no longer operate the joystick, other options like head controls or switches that are operated by other parts of the body, are possible.

Due to the complexity, these types of wheelchairs are very expensive and financial resources like insurance coverage, not-for-profit organizations and provincial programs should be considered. It is therefore imperative that a power wheelchair be assessed, trialed and prescribed by an occupational therapist with expertise in these types of wheelchairs.

Lifts and Stair Glides

This type of equipment is used to address home accessibility problems or to make it safer and easier for you to transfer from one surface to another. It can be expensive and requires much consideration and preplanning to determine what is the best solution for your home-setting and longer-term situation. The types of lifting equipment to consider include lift chairs, stair glides, portable lifts, ceiling-track lifts and wheelchair lifts. **It is best to get an occupational therapist to assess your residence and situation when making your plans regarding lifting devices.**

- **Lift Chair**

People with weakened legs will find it difficult to get up out of chairs. Higher chairs with arms to push up can make it easier. Ideally, a **motorized lift chair can be used to safely raise a person from a seated to a stand.** In addition, lift chairs also recline to optimize sitting posture and comfort and to allow gravity assisted positioning for people with a weak neck and/or back. Some models come with heaters, a massage device and other features that may be beneficial.

- **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 that attaches to the ceiling 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 Floor Lifts**

Portable floor lifts also use a sling to lift a person up. They can be rolled around on one level of the home, but require sufficient floor space to manouver. A common type of floor lift is the **Hoyer Lift.** Some of these kinds of lifts are light, can be broken down into two pieces, and moved to a different level of the home, or put in a car. The minimum door size for moving a person through in a portable lift is about 26". Using a ceiling or floor lift requires training, to ensure the safety of both you and your caregiver.

- **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, may not be a good long term solution, as safe sitting balance and ability to transfer on and off is required. **Stair glides should only be installed by a professional familiar with the safety aspects of this equipment.**

- **Porch Lifts**

Porch lifts are often used when it is not practical to have a wheelchair ramp from an outside door to ground level. Again, **installing a porch lift system is a major project which requires advice and installation by a professional who is familiar with all safety aspects of installation and use.**

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 homemade 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 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 **bathtub 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 the right equipment for you.

Beds and Mattresses

A common problem during the advanced stages of ALS is being unable to roll over in bed. Lying in one position can become painful, or a caregiver is required to move you every few hours. **Satin sheets** and or pyjamas can make turning easier. There are also different mattresses and overlays of various materials like foam, gel, and air which can optimize comfort and pressure relief.

A bed with an incline feature can be helpful with breathing and comfort. Devices from **foam wedges** to fully automatic hospital beds are used.

Augmentative and Alternative Communication (AAC) Equipment

There are many different types of assistive devices available to help communicate if speech is impaired. These devices range from alphabet and message **boards**, 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.** Your choice will depend on your specific needs, available resources 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
- **Wheelchair model** can attach to your wheelchair allowing you to bring it wherever you need to go
- **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 slope equals 12 inches length for every one inch height) and guardrails
- **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. 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** should be completely barrier free with no step 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.**

SERVICE DOGS

In recent years, service dogs have become an assistance option (and companion) for people with ALS. They can be taught various tasks, such as carrying or fetching objects and opening doors. They can be trained to pull a wheelchair or brace a person so they can independently transfer from one seat to another. Service dogs can also be trained to bark for help, alert a family member or caregiver in the event of an emergency and lie down on its partner's chest to produce a cough. Several organizations across Canada train service dogs for people with ALS. Contact your Provincial ALS Society for a list of trainers.

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 diagnosis of ALS may result in increased expenses. This may be true, whether one remains at home, or if one resides in a Long Term Care Facility. Caregiving support, equipment, medical supplies and medications for symptom management are just some of the costs associated with a diagnosis of ALS. These additional expenses may not be covered by provincial programs, extended health benefits or non-profit organizations and therefore become the responsibility of the client and their family. In some provinces, there are support and funding resources available to assist with these new expenses. It is important to check with the local ALS Society and healthcare team around what supports are offered and how to access them. **The assistance provided by the ALS Society becomes, for many people, a critical factor maintaining quality of life.**

EMPLOYMENT AND ALS

For those who have been diagnosed with ALS and continue to work, one of the things to consider is when and what to tell your employer. This is a personal decision depending on your and your employment situation.

In light of a diagnosis of ALS, you may wish to consider exploring any extended healthcare **benefits** you might be eligible for. It is important to consider your current needs in addition to what may be required in the future, as there may be limits to what is provided. Consultation with your healthcare team is recommended while investigating these resources. **All of this can be done before telling your employer you have ALS.**

When Do You Tell Your Employer?

Here are some factors to consider before notifying your employer:

- Are you working on a project that you want to finish, before telling your employer?
- Are your symptoms interfering with your ability to perform your job?
- Are you concerned that your employment future would be compromised if you told your employer?
- In Canada it is a legal requirement to "accommodate" a person with a disability such as ALS. If you have questions about this policy please contact your ALS team social worker or consult an attorney specializing in employment law.

Factors to consider before leaving work:
- Do you have access to benefits?
    - Private Pension, Extended Health Care, Short Term Disability, Long Term Disability, Banked Sick Time, Medical EI, Canada Pension Plan etc?
- Does working bring meaning and quality to your life that would be missing if you were unable to continue working?
- Have you made modifications, but are still unable to carry out your job duties?
- Do you have the energy to continue working?
- What financial supports do you have? (RRSPs, Savings, Investments, etc.)

It is highly recommended that you do not sign anything regarding your termination without first getting legal advice. You may be eligible for serance packages and benefits.

Should you have any questions about what goverment programs and benefits you may be eligible for, please contact your social worker.

What Protections Do You Have Under Provincial Labour Legislation?

The legislation for each province varies with respect to employee rights. Consider getting advice from a lawyer who specializes in human rights, provincial employment standards and labour legislation. Many provinces have free or subsidized legal support available to residents. Consult with your ALS Clinic social worker who may be able to answer some questions around relevant legislation and provide information on local resources.

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 providing 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.

Below is a guide that may be helpful when contacting your insurance company. It is advised that you have all of the specific information about your insurance plan in front of you while calling. Feel free to ask what certain words or terms mean.

Group insurance can be valuable resource. If you have group insurance through your employer, you may need to seek advice from a lawyer union representative, or someone else you can trust to ensure that this insurance coverage continues to be effective when you are. Should you experience difficulties around coverage, you may wish to consult an attorney about your right.

Driving and Property Insurance

Driving can sometimes require fast foot and hand reactions. Speak with your doctor about whether you are safe to continue driving. A driving test may be required to verify your current abilities. Please speak with your local health care provider about how to access these services. Also, note that the legislation pertaining to driving safety varies from province to province. Should you have any questions about this, pleas e contact your local motor vehicles department.

If you have specialized equipment, have made modifications or renovations to your home, as the result of your diagnosis, you may need additional property insurance. Review your current plan and speak with an insurance representative to ensure coverage or if you have any questions.

Life Insurance: Living Benefits

Some life insurance companies offer a "living benefit" feature to people with a terminal disease enabling them to recieve a portion of their life insurance paid to them in advance, during the years before their death.

- **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 accountant 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.

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?

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 as all polcies are not the same. 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 such organizations.

PENSION PLANS

Group Pension Plans

If you are in a company or other group pension plan, find out about any changes to your plan 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.

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 under 65 years of age
- Stopped working because of your medical condition
- Made enough contributions in four of the last six years you were employed.

Or

- Have paid into CPP for at least 25 years and made valid contributions to the Plan in three of the last six years.

- **What if I am already retired and receiving CPP?**

If you are receiving a CPP retirement pension you can apply to have your retirement pension replaced by a CPP disability benefit if you became disabled (according to CPP legislation):

1. before you turned 65;
2. before your retirement pension began.

In addition, your application for CPP disability benefits must be made within 15 months of the start of your CPP retirement pension. Any CPP retirement pension payments you have already received may be deducted from your disability benefit.

Conditions of Eligibility

There are two conditions that you must meet:

Definition of disability

1. The disability must be considered to be severe and prolonged as defined under CPP legislation.

CPP contributions

2. You have to have made enough contribu tions into the CPP while you were working. (see below)

(Source: "Service Canada" website)

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.

Compassionate Care Benefits for Caregivers

Eligible Canadian workers who take time off work to care for a gravely ill family member can receive up to six weeks of employment insurance benefits over a period of six months and will have their jobs protected.

To qualify for compassionate care employment insurance benefits, applicants must have worked 600 hours in the previous 52 weeks and show that regular weekly earnings have decreased by 40%. They must also provide a medical certificate to show that the family member has a high risk of death within 26 weeks. Compassionate care includes providing emotional support, arranging for third-party support or directly providing the care. Benefits may be shared among eligible family members.

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

Effective in 2006, the definition of family member was expanded to include those who provide care for a brother, sister, grandparent, grandchild, in-law, aunt, uncle, niece, nephew, foster parent, ward, guardian, or a gravely ill person, such as a neighbour or close friend, who considers the claimant to be like a family member. Benefits remain available to those caring for a parent, child, spouse or common-law partner. Note that common-law partner means a person who has been living in a conjugal relationship with that person for at least one year.

Compassionate care benefits can be paid even if the ill family member does not live in Canada. For additional information visit www.hrsdc.gc.ca/en/ei/types/compassionate_care.shtml.

TAX ISSUES

There may be a number of ways to more effectively manage your tax situation resulting in greater after-tax income. Talk to your accountant and/or financial advisor about your options.

Disability Tax Credit

The Disability Tax Credit is a non refundable credit for eligible individuals or their representatives. It reduces income tax payable on your income tax return. Eligibility requirements are outlined on the application (T2201) which is available on the Revenue Canada website. The application must be completed and signed by your doctor. It is advised that you send your application in prior to tax season to avoid delays. Application processing times vary. Please keep a copy for your records.

Other Tax Credits:

- Caregiver Tax Credit
- Attendant Care Expense Deduction
- Fuel Rebate (provincial and federal)
- Medical Expense Tax Credit

You may also be eligible to claim certain medical expenses. Information on eligibility for tax programs can be found on the Revenue Canada website: http://www.cra-arc.gc.ca/disability/. Your financial advisor, accountant, or social worker can assist you

with the process of applying for any such benefits.

Minimize Tax Through Income Splitting

Income splitting reduces a family's total income tax by dividing income among various family members, using the low tax rates of family members who have little other income to offset the higher tax rates of those with higher 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

This list is not exclusive. If substantial funds are involved, or even if funds are minimal, see a professional tax expert who specializes in disability issues.

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 decreased 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 required to pay U.S. taxes on the estate.

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.

Review your Will and Powers of Attorney to ensure that these reflect your current wishes. Keep reading for more information about living wills and powers of attorney.

FUTURE PLANNING DOCUMENTS

Power of Attorney

A Power of Attorney is a document that allows an adult to appoint an individual to make decisions regarding their financial and legal affairs on their behalf. This "attorney" is authorized to act when the adult becomes incapable, due to limited mental ability or illness. You may choose one or more persons to act

as your power of attorney. If you select more than one person, you must indicate whether any named individual can make decisions for you alone, or whether they must make decisions as a group.

The Powers associated with a Power of Attorney vary from province to province. In some provinces, this document gives the "attorney" the authority to make decisions around your health care. Please check with your clinic social worker about relevant provincial legislation.

HEALTH CARE DECISION MAKING

Power of Attorney for Health Care

In some provinces, a Power of Attorney document can be used to appoint an "attorney" who can make health care decisions your behalf should you be unable to. This is called a Power of Attorney for Health Care (PAHC). Your Power of Attorney for Health Care has the legal and ethical obligation to make decisions about your health care that respect your wishes. Therefore, it is important to have open discussions around what those wishes might be.

Representation Agreements

In other provinces, alternate decision makers are appointed through what are known as Representation Agreements. Representation Agreements are legal documents that appoint another individual to make healthcare decisions on your behalf, should you be unable to. There are two parts to a Representation Agreement; each providing different powers.

Section 7: Allows you to name an alternate decision maker who will have the authority to make decisions around routine management of your financial affairs, your personal care and some health care treatment. It does not allow for your representative to make health care treatment decisions that involve life sustaining or life ending interventions.

Section 9: Allows you to appoint a representative to make decisions on your behalf around personal care and health care treatments; including decisions pertaining to life sustaining and life ending interventions. A Section 9 Representation Agreement does not allow for your representative to make decisions about your financial matters.

Temporary Substitute Decision Making

In some provinces, in the event you have not legally appointed an alternate health care decision maker and you require someone to make such decisions on your behalf, a Temporary Substitute Decision Maker (TSDM) will be appointed by your healthcare team. A Temporary Substitute Decision Maker is chosen from a predetermined list and must meet certain criteria. The TSDM is obligated to make decisions that are in line with your wishes. The legislation surrounding TSDM varies from province to province. If you are uncertain as to who your TSDM will be, please consult with your healthcare providers.

Informing Your Alternate Decision Maker

There are numerous ways in which you can inform your family, Power of Attorney for Health Care, Representative or TSDM. Below please find a summary of each.

LIVING WILLS AND ADVANCE CARE PLANS

A Living Will, or Advance Care Plan, is a document that outlines your beliefs, values and wishes regarding future healthcare treatment. A living will or advance care plan is used by your alternate decision maker and your healthcare team as a guide to understanding what your wishes around future interventions might be should you be unable to communicate them yourself. This is a general guide about your wishes and may not cover every possible outcome. That is okay. It will be up to your medical team and your alternate decision maker to determine the most appropriate course of action that is in line with your values and beliefs. This is why it is important to share your values and beliefs with your healthcare team and your alternate decision maker. **Below are some things to consider when writing your living will or advance care plan:**

- What makes my life meaningful?
- What and who do I care about in my life?
- How do I define quality in my life?
- What medical interventions would help maintain quality of my life?

i.e. would I want life sustaining interventions such as CPR, Life Support? Or would I prefer a comfort care approach to end of life?
Remember, an advance care plan or living will only comes into effect if you are unable to communicate your wishes to your healthcare team.

Once again, legislation in each province varies, as does terminology. If you have any questions about how or when a living will or advance care plan is used, or would like help creating one, please speak with your healthcare provider.

Advance Care Directives

In some provinces one can create a legally binding document that acts as direct communication to your doctor and healthcare team. These are sometimes known as Advanced Directives. These directives, outlining your wishes, act as instructions to your healthcare team. The directive only comes into effect if it relates to the current medical situation and you are unable to reliably communicate your wishes. The directive may take legal precedence over your alternate decision maker as it is considered direct communication of your wishes.

- **Why should I complete these documents?**

There are benefits to having these conversations and completing this documentation. For instance, these documents provide an opportunity for you to express your thoughts and wishes. They also help to reduce the stress your family and an alternate decision maker may experience in the event they have to make a decision on your behalf. **It is advised that you begin this process in the earlier stages of your disease when you are relatively healthy. Please share a copy of these documents with someone you trust and your healthcare providers.**
As a person with ALS, you will experience many changes in health and functional status, and as such, your perceived quality of life may also change. Therefore, what you decide you want today, may change a year from now. **You may change your living will, advance care plan or advance directive at any time. It is advisable to revisit your planning documentation often to ensure that it accurately reflects your wishes. In the event you decide to change your plan** please ensure that all pre-existing versions are destroyed. It is also important to notify those involved in your care of any changes to your plan and redistribute the revised plan accordingly.

Depending on which province you live in, you may need to consult with an attorney in order to create some of these documents. If you are unsure how to proceed, please contact your healthcare provider.

- **Where Can I get Sample Advance Directive Documents?**

Advanced Care Planning documents are often available at no charge through your **provincial government** and can be downloaded on-line. A sample healthcare directive created specifically for palliative care patients, is available on-line at www.palliative.info, and 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). This form is a figure contained in:
Benditt, Smith, and Tonelli (2001). Issues and Opinions: Advance Care Planning in ALS. Muscle and Nerve, **24**: 1706-1709.

Contact your ALS Society office for a copy of this form.

ESTATE PLANNING

Estate planning is the process by which you begin to get your financial affairs in order. Estate planning can include the creation of a Will, examination of assets and liabilities and the setting up of Trusts. Estate planning may reduce the amount of probate fees and taxes that your estate would otherwise pay. Estate Planning also gives you an opportunity to express your wishes for once you have passed away.

Having an Estate Plan may anticipate different scenarios, but it is important to think about updating it whenever your financial or personal circumstances change. You may also want to change your Estate Plan if there is a change in the beneficiaries.

An Estate Plan may also document what your wishes are for once you have passed away. This can include any spiritual or religious service you may wish to have

in your honour. Having an Estate Plan provides guidance to your family during a time of great loss.
Below please find some additional information on creating a Will and some considerations for when planning your Estate.

LAST WILL AND TESTAMENT

A Will is a legal document that provides instructions to your loved ones on how some of your assets and liabilities (i.e. property, investments, money) are distributed once you have passed. A Will may minimize the additional income taxes, and probate fees payable upon your death. It may also minimize taxes payable by your beneficiaries on future income. A Will may not cover all assets and investments. As a result, it is important to plan your Estate in order to ensure that your wishes are met with minimal impact on your beneficiaries.

Planning A Will

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.
• If you have minor children, appoint a Guardian in your Will.
• Consider if you want to appoint a Guardian of the Estate. This guardian can receive funds from your executor for the benefit of your children. The person you choose may be the same guardian you choose for your children or someone different.
• 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

Making A Will—Formal Requirements

A Will is only valid if it meets the requirements set out in the Wills legislation in the province or territory in which you live. Generally, the normalities include dating the Will and having the maker of the Will (the "Testator") sign it in the presence of two witnesses, who should not be beneficiaries under the Will. The witnesses must also sign the document in the presenceof the Testator and each other.

To avoid possible confusion in the future, only one copy of a Will should be signed. Many provinces also allow for the preparation of a "holograph" Will, which is a document that is completely in the handwriting of the Testator and is dated and signed by the Testator. However, because of many drafting and other problems, holograph Wills are often a source of conflict and litigation, so their use should be discouraged.

For similar reasons, the use of Will kits is not recommended. Given the broad range of legal areas and issues that must be considered in
planning and preparing a Will, some of which are outlined here, there is really no substitute for the professional legal advice of a lawyer
whose practice is focussed on will, estate and tax planning.

You must also have the appropriate mental capacity to make a Will and must sign it willingly, without duress or improper influence by other individuals. Otherwise, the Will can be challenged and would be of no effect. For this reason, it is important to have a Will prepared while you are competent and capable, to
ensure that your intended beneficiaries will benefit as you direct. It is also generally recommended that you review your Will.

Appointment of Executors

An important advantage of making a Will is that it enables you to decide who will be the "executor" who will administer your estate.
The executor of the estate plays an important role, since the executor is responsible for many tasks, which include collecting the assets of the estate, paying taxes and any other liabilities, and distributing the assets according to the
terms of the Will.

Given the significance of an executor's duties and obligations, the selection of the executor should be carefully considered. When choosing an executor, you should consider certain factors, including:

• The complexity of your assets and your estate

• How your estate is to be distributed. A Will leaving only outright gifts to intended beneficiaries will generally be less complex to administer than an estate under which trusts are to be established for one or more beneficiaries

• The age of the proposed executor. It is generally preferable to name an executor who is younger than you, as this makes it more likely that your executor will be available and able to act when and for as long as required
whether a proposed executor has the necessary skills and abilities to administer your estate effectively

• Where the proposed executor lives. Executors who do not reside in Canada have to post a bond as security before being allowed to administer an estate and can create considerable complexity for the estate and for the beneficiaries since they may also cause the estate to be non-resident for tax purposes

• The trustworthiness of the proposed executor.

It is important to review your Will every few years to make sure that it continues to reflect your wishes.

If one does not have a Will, your assets and liabilities will be divided according to relevant legislation and this may not happen the way you would have wanted. **Should you have questions about how to prepare an Estate Plan, please consult with a lawyer or estate planner.**

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.

family member or friend to help get you organized or do research on your health benefits and legal rights can make it more manageable.

Final Note on Financial and Legal 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.

NOT TOO BIG. NOT TOO SMALL.

Pallett Valo has the depth and breadth of experience to assist you with the challenges you and your family may face, while still delivering personal and compassionate service at realistic and affordable rates.

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- Estate Litigation
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- Workplace and Accommodation Issues
- Guardianships

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After his ALS diagnosis in April 2005, Pallett Valo LLP founding partner Sidney Valo resolved to accept his condition as his "new normal" and do all he could to help others afflicted with ALS including joining the board of the ALS Society of Canada and establishing the Valo Fund for ALS Research.

We were privileged to have had Sid Valo as our partner and friend. Sid will be remembered by all of us at Pallett Valo LLP.

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

Dementia and Motor Neuron Disease
Edited by Michael J. Strong
Informa Healthcare (2006)
This book is the single authoritative reference on the current understanding of frontotemporal dementia in ALS.
ISBN: 0415391660

Motor Neuron Disorders
Edited by:
Pamela J. Shaw, MD
Michael J. Strong, MD
Butterworth-Heineman/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
By Robert G. Miller, MD, Deborah Gelinas, MD, & Patty O'Connor, RN.
This book is one of the first in a serious 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, 3rd Edition
Edited by Hiroshi Mitsumoto, 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, Suite 301
New York, NY 10016
ISBN:1932603727
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

Breathe Easy: Respiratory Care in Neuromuscular Disorders (2009)
This book can be downloaded at www.mda.org/publications/PDFs/BreatheEasy/Booklet.pdf. It covers respiratory management options, equipment, etc. in ALS as well as muscular dystrophy.
MDA-ALS Division
3300 East Sunrise Drive
Tuscon, AZ 85718-3299

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

Everyday Life with ALS: A Practical Guide
Produced by MDA-ALS Division, it is a practical guide that answers many questions about daily living, equipment, therapies, and accessibility that arise over the course of the disease. It includes many helpful photos and diagrams. It can be downloaded at www.als-mda.org/publications/everydaylifeals/ or for $15 USD can be purchased by contacting (800) 572-1717 or publications@mdausa.org.
MDA-ALS Division
3300 East Sunrise Drive
Tuscon, AZ 85718-3299

Guide to ALS Patient Care for Primary Care Physicians (CD-ROM)
Produced by the ALS Society of Canada, this resource is designed to inform primary care doctors who have patients with ALS to ensure continuation of quality care in the community between visits to ALS specialists. A person with ALS who receives care at an ALS specialty clinic can request a CD-ROM be sent to their family doctor. For patients who do not attend ALS specialty clinics, a copy of this resource can be obtained from the local Provincial ALS Society, or by downloading it at http://www.als.ca/publications-and-resources/physician-cd

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

Motor Neuron Disease: A Family Affair
By David Oliver, MD
Sheldon Press
36 Causston St.
London SWIP 4ST
ISBN: 0859699773

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: 158182348

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

A Caregiver's Guide: A Handbook about end-of-life care
This book provides family and inforamtl caregivers te medical and nursing information they need in easily understood language. This guide will also help build a strong palliative care team who are effective in providing physical, spiritual and emotional support.
Published by the Canadian Hospice and Palliative Care Association (CHPCA) and athe Military and Hospitaller Order of St. Lazarus of Jerusalem. To order, visit http://www.chpca.net/.

A Guide to Caregiver, You Are Not Alone Living Lessons®
1-877-237-4363 (INFO), or download from www.living-lessons.org

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 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
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, clearsighted, 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

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 hat 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
http://www.rgp.toronto.on.ca/

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

Influencing Change: A Patients and Caregiver Advocacy Guide
By the Canadian Hospice and Palliative Care Association (CHPCA)
This book is part of the Living Lessons® project of the CHPCA and hospice and palliative care organizations across Canada. The book provides readers with the tools and information to help them become healthcare champions for themselves or a loved one.
1-877-203-4636
www.living–lessons.org

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.
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

Our Greatest Gift: A Meditation on Dying and Caring
By Henry J.M. Nouwen
Reflecting on his own impending death, as well as on the deaths of loved ones and friends, Nouwen shares his view of death as a gift, an opportunity to impart hope to hose around us, whether through our own dying or in our caretaking of someone else.
Harper One
ISBN: 0060663553

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

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 Suffering Persists
By Ira Byock, MD
Reflections on suffering and commitment to alleviate it by an American hospice physician.
Journal of Palliative Care, 10:2, pp. 8-13
Also available at http://www.dyingwell.org/suffper.htm

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

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 o 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 Donelly, USAF, Retired
With Denise Donelly
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/
IBSN: 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
ISNB: 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

I Remember the Year I got Everything I Wanted –And ALS
By Darcy Wakefield
Marlowe and Company
245 West 17th St.
New York, NY 10011
ISBN: 1569242798

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

ALS Activity Book
Produced by The ALS Association, St. Louis Regional Chapter
2258 Weldon Pkwy.
St. Louis, MO 63146
(888) 873-8539
www.alsa-stl.org
$10 USD

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

Dear Bradie: A Story of Life with ALS
By Martha Brunell
An audiobook of 45 letters written for three year old Bradie, by her father Jim about what it's like to live and die of ALS at 40.
Contact Martha Brunellat mbrunell@access.net to order or visit www.wildwithwords.com/bradie.htm

Grandpa, What is ALS?
ALS Society of Alberta
400, 320-23 Avenue S.W.
Calgary, AB T2S 0J2
(403) 228-7752

The Grieving Child: A Parent's Guide
By Helen Fitzgerald
Simon and Schuster
New York, NY
ISBN: 0671767623

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

Helping Children Cope with ALS—A Parental Information Guide
Produced by the ALS Society of Canada. To request a copy, contact your Provincial ALS Society, or download for free at www.als411.ca.

How to Help Children Through a Parent's Serious Illness
By Kathleen McCue
St. Martin's Press, New York
ISBN: 0312113501

In My Dreams...I Do!
By Linda Saran
Inspired by the author's mother who continued to nur-

ture 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

Lifetimes
By Bryan Mellonie and Ropbert Ingpen
Excellent book about the life cycle for younger children.
Bantam Books, New York
ISBN: 0553344021

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.

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
(612) 926-2665
ISBN:0962050242

Patrick and the Giant
By David G. McNelis
When Patrick, the village storyteller, hears his name being called out by a thirty foot tall giant, he is terrified. Using humor to hide his fear, Patrick confronts the giant and discovers that this hideous creature has cursed him with an evil sickness, and has only days to live. With his body quickly weakening, from the giant's illness, Patrick decides he must slay the monster in order to survive. Aided by his family and Connor, the red bearded blacksmith, the battles begin. It soon becomes apparent that destroying a mighty giant is no easy task. However, Patrick refuses to give up which all makes for a tale full of exciting confrontations, unexpected twists, and plenty of laughs and tears along the way.

David McNelis worked as a Physician Assistant for over twenty years. In 1996, he was diagnosed with ALS. Having to retire in 2002, he began writing humorous stories about his childhood, as a legacy for his children. Patrick and the Giant is his latest endeavor, which he managed to complete with his last working finger. He continues to write with the use of his foot. He and his wife Linda live in New Jersey and have four children and three grandsons.
Wasteland Press
www.wastelandpress.net
ISBN: 978-1600474910

Saying Goodbye: Bereavement Activity Book
By Jim & Joan Boulden
Boulden Publishing
P.O. Box 9249
Santa Rosa, CA 95405
(800) 238-8433
ISBN: 1878076124

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

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

Talking With Young People About ALS: For Schools
Produced by the ALS Society of Canada. To request a copy, contact your Provincial ALS Society, or download for free at www.als411.ca

Talking With Your Patient's Children About ALS: A Primer for Health Professionals
Produced by the ALS Society of Canada. To request a copy, contact your Provincial ALS Society, or download for free at www.als411.ca

The Grieving Child: A Parent's Guide
By Helen Fitzgerald
Simon and Schuster, New York
ISBN: 0671767623

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

When Someone Special Has ALS—A Booklet for Children
Produced by the ALS Society of Canada. To request a copy, contact your Provincial ALS Society or download for free at www.als411.ca

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

When Your Parent has ALS—A Booklet for Teens
Produced by the ALS Society of Canada. To request a copy, contact your Provincial ALS Society, or download for free at www.als411.ca

VIDEOS/DVDs

ABC News Presents Morrie Schwartz: Lessons on Living
Tuesday's with Morrie proved to be a highly popular memoir, in which college professor Morrie Schwartz was caught in conversation with former student Mitch Albom. Albom was inspired to write this book after catching Morrie in conversation with Ted Koppel, on his Nightline television show. Attempting to chronicle his dying days on Keppel's show, Morrie's recollections are earnest, heartwarming, and a fascinating glimpse into one man's final moments on the planet. They are presented here, and are both funny and sad in equal measure, with Morrie displaying all his trademark wit despite his failing health due to ALS. See bookstores and on-line resources such as www.cuniverse.com, for a copy of this DVD.

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 indepth, 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

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.

Hospice Foundation of America Videos:
(800) 554-3402
www.hospicefoundation.org

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 practical 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: 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: 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 grievers 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.

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.

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.

Nutritional Lifeline: The Feeding Tube Decision in ALS
www.simplifiedtraining.com
Contact your Provincial Society to request a loan from their lending library.

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

So Much So Fast
What would you do if you were 29 and found you may only have a few years to live? So Much So Fast is about the remarkable events set in motion when Stephen Heywood discovers he has ALS and his brother Jamie becomes obsessed with finding a cure.
West City Films, Inc.
To order: http://westcityfilms.com
$26.95 USD

State of the Art Communication for PALS
Produced by www.synvision.dk
For more information: www.als-communication.dk

The Man Who Learned to Fall
Produced by Beitlel/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).

INTERNET RESOURCES

ALS Specific

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 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 Society of Manitoba
- www.envision.ca/webs/alsnl
  - ALS Society of Newfoundland and Labrador
- www.alsnb.ca
  - ALS Society of New Brunswick
- www.alsns.ca
  - ALS Society of Nova Scotia
- 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.lesturnerals.org
  The Les Turner ALS Foundation (Chicago-based ALS organization)
- www.march-of-faces.org
  March of Faces - raising awareness project
- www.als-mda.org
  Muscular Dystrophy Association (MDA) (US) ALS Division supports people with ALS and funds ALS research
- www.mnda.org
  Motor Neurone Disease Association (UK)

Resources for Children:

- www.als411.ca
  ALS Canada web site for children and teens
- www.march-of-faces.org/KIDS/moe1.html
  March of Faces space for kids
- www.alsindependence.com

Primary Lateral Sclerosis - PLS:

- www.geocites.com/freyerse
  - maintained by an individual with PLS

Basic Information - Sponsored by a Research Organization:

- www.nlm.nih.gov/medlineplus/amyotrophiclater alsclerosis.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.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
- www.outcomes.umassmed.org/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

About Rilutek - manufacturer site -

- http://www.sanofi-aventis.ca/products/en/rilutek.pdf

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

http://www.ltcplanningnetwork.com/
How To Care

- www.chpca.net
  Canadian Hospice and Palliative Care Association (CHPCA)
- 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 sup portive 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 care givers through experts, articles, newsletter, jour nal 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 feder ally 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://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 vari ety 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 pro grams for individuals. Of particular interest may be the pages for seniors, people with dis abilities 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 organiza tions and international health information providers. The resources identified here will help you take care or 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 other wise 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.

FACT SHEETS

Fact Sheets from the ALS Society of Canada, can be printed from the web site www.als.ca and stored in the

pocket page for your reference.

New Fact Sheets are published periodically, so check the web site for additional topics to those listed below at www.als.ca/publications-and-resources/als-fact-sheets.

#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

RESEARCH UPDATES

The Research Updates referred to in the Manual can be downloaded from http://www.als.ca/en/research/our-research-program and stored in the pocket page for your reference. Please check web site link for additional topics, including those below.

#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