



# PREFACE

The purpose of this electronic publication is to provide primary care physicians and other healthcare professionals information about amyotrophic lateral sclerosis (ALS). We hope this guide will assist you to:

- recognize signs and symptoms of ALS and make timely referrals to ALS specialists
- understand ALS progression, symptom management, changing patient needs, and the importance of interdisciplinary ALS care
- help your patients and caregivers cope with living with this very complex and progressive neuromuscular disease

Ideally, the care of ALS patients and caregivers is best provided by the collaboration of an informed family physician, a specialty ALS team, homecare providers, and the local ALS Society. Specialty ALS centres are staffed by an interdisciplinary healthcare team, under the direction of a neurologist or physiatrist who specializes in ALS. ALS clinics and centres ideally monitor patients every three months to assess progression, adjust care plans, and make recommendations to maximize quality of life. However, patients with ALS may seek help to manage symptoms and discuss coping with the disease with their family doctor between visits, and in some cases the family doctor will need to play the lead role in cases where the patient chooses not to attend a specialty clinic after diagnosis due to geographic barriers, problems with traveling, or personal preference. Over the course of the disease, ALS patients will need homecare at increasing levels, assistive devices, and other community-based services, so a well-informed primary care practitioner who can help advocate for an ALS patient is critical.

In addition to this Guide, we encourage you to read the current *American Academy of Neurology (AAN) Evidence-Based Guidelines in ALS* published in 2009 as an update to the original ALS Practice Parameter in 1999. The two full articles can be accessed through the following link:

<http://www.aan.com/practice/guideline/index.cfm?fuseaction=home.welcome&Topics=19&keywords=&Submit=Search+Guidelines>

Summaries of the Evidence-Based Guidelines for Clinicians and Patients (four PDF files in total) and a PowerPoint presentation on the development of the Guidelines can be accessed by going to [http://www.als.ca/als\\_care\\_guidelines\\_professionals.aspx](http://www.als.ca/als_care_guidelines_professionals.aspx).

Please also visit the web site of the ALS Society of Canada ([www.als.ca](http://www.als.ca)) periodically for updates on research and professional and patient resources in both French and English.

Bottom line, it is essential for any healthcare provider in the community with an ALS patient to understand this devastating disease and its physical, emotional, mental, and financial implications to ensure the best possible care and support.

# ACKNOWLEDGEMENTS

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# INTRODUCTION TO ALS

**A**myotrophic lateral sclerosis (ALS), commonly called Lou Gehrig's disease or Motor Neuron Disease, is a relentless, fatal neuromuscular disease. ALS is marked by **progressive and highly selective degeneration and loss of upper and lower motor neurons in the brain and spinal cord leading to paralysis of voluntary muscles and loss of ability to swallow, speak, and breathe.** Until recently it was believed that ALS did not affect cognitive functioning. Recent studies have shown that mild forms of cognitive impairment (e.g., executive function, insight) and behavioural changes (e.g., compulsive ritual-like behaviour not previously exhibited) may occur in as many as 50% of ALS patients. Florid dementia is rarely reported. More research is needed in this area to better understand the pathology and treatment of ALS.

The **region of onset** (limb versus bulbar) varies, but in the majority of cases it presents as limb onset ALS. The rate of progression varies from one individual to another, although it generally remains constant. ALS is neither contagious, nor infectious.

## CLASSIFICATION

Epidemiological and genetic factors permit classifying ALS as:

- **Classical** sporadic ALS (90% of cases)
- **Familial** ALS [5-10% of cases in which 20% of these are connected to a mutation of copper zinc superoxide dismutase 1 (SOD1) in chromosome 21]

These syndromes are associated with a loss of cortical motor neurons, anterior horn cells, and bulbar motor neurons, as well as with secondary gliosis in the corticospinal tract.

**NOTE:** Other motor neuron disorders such as progressive muscular atrophy (PMA), progressive bulbar atrophy, primary lateral sclerosis (PLS), and Kennedy's disease share some of the signs and symptoms of ALS.

## DIAGNOSIS

Diagnosis is made by a neurologist. Every person with suspected ALS should be sent to an ALS special-

ty clinic for assessment and a confirming diagnosis. As yet there is no specific laboratory test for ALS; diagnosis is supported by neurological and clinical investigations. There are **three principal diagnostic factors:**

- The presence of progressive neuromuscular signs and symptoms, usually described as weakness, muscle wasting, stiffness, muscle twitching (fasciculations or involuntary muscle contractions), or muscle cramps, although some patients will report "numbness" of a weak or paretic area. (**NOTE:** Although usually not seen until later stages, some patients may present with acute respiratory symptoms.)
- The results of investigations with electromyography (EMG), possibly MRI, and blood tests
- The reasonable exclusion of all other disorders that may mimic signs and symptoms of ALS

Insidious onset of weakness may not be initially recognized as ALS, and even when suspected, confirmation of the diagnosis may take several months to a year. However, a diagnosis of probable or definite ALS confirmed by an ALS specialist, is seldom incorrect, in part because of the methodical diagnostic process. Giving a patient a diagnosis of ALS requires great sensitivity and compassion. A step-wise disclosure while investigations are underway, may ease acceptance once the diagnosis is confirmed.

## CLINICAL SIGNS AND SYMPTOMS

ALS usually begins **focally** with mild weakness and muscle wasting or spasticity. The initial involvement is **asymmetric**, but muscle wasting and weakness gradually become bilateral and widespread. Impaired function may be first seen in the muscles of the hand or leg, or with changes in speech or swallowing. In a minority of ALS patients the loss of motor function that is the hallmark of the disease can be accompanied by marked alteration in reasoning and social interaction that can further disable a patient and burden families already under enormous stress. However, behaviour effects are often mild, or indeed absent. Bowel and bladder control usually remain normal and the sensory system is seldom impaired.

# INTRODUCTION TO ALS

Signs of **lower motor neuron degeneration** occur in all cases of ALS as the disease progresses. They are as follows:

- Muscle weakness, atrophy (including diaphragm)
- Fasciculations
- Muscle cramps
- Hyporeflexia
- Flaccidity

Signs of **upper motor neuron degeneration** (mild spasticity with hyperreflexia and a Babinski sign) are present in 85% of cases. Other signs and symptoms may include the following:

- Contractures
- Dysarthria (a speech disorder caused by impairment of the muscles used for speaking)
- Dysphagia (difficulty in swallowing)
- Dyspnea (shortness of breath at rest)
- Emotional lability (uncontrollable, often inappropriately timed, laughing or crying)
- Excessive fatigue
- Sialorrhea (excessive salivation)
- Weight loss

The presence of upper and lower motor neuron signs is required for the diagnosis of ALS. One or the other may predominate in an individual patient, and may even vary between regions in the same patient. One limb may be spastic with hyperreflexia, another flaccid and atrophied with hyporeflexia.

## AGE, INCIDENCE, PREVALENCE AND MORTALITY

The average age of diagnosis of ALS is 55 years; **most cases are diagnosed in individuals between 40 and 70 years old.** However, ALS has occurred in teenagers, and may be found in the oldest old (over 90). The estimated crude incidence rate based on total population is **2/100,000 per year**; the estimated prevalence is **6-8/100,000 on any given date.** However, if only considering populations over 20 years of age the incidence is more likely 3-4/100,000. National incidence rates are also influenced by age composition, i.e., a population with a much greater proportion of young people will likely see a lower incidence rate than one with an older population. Life expectancy is typically 2-5 years while some die sooner and other live much longer, i.e., 20% over 5 years and 10% over 10 years.

## RELATIONSHIP OF SYMPTOMS TO LOCATION OF MOTOR NEURON LOSS

Medulla	Upper Motor Neuron Lesions	Pseudo Bulbar (other causes-including stroke)	Tongue spastic Speech spastic, explosive & slurred (dysarthria) Increased reflexes Emotional lability Incoordination of respiratory and swallowing functions
	Upper and Lower Motor Neuron Lesions		Dysarthria Dysphagia Wasting of tongue Jaw jerk reflex increased
	Lower Motor Neuron Lesions	Bulbar Palsy	Tongue-atrophied, corrugated, fasciculating Speech slurred Dysphagia
Cortico Spinal Tract	Upper Motor Neuron Lesions		Spastic weakness Stiffness Increased reflexes Extensor plantar responses
Anterior Horn Cells	Lower Motor Neuron Lesions		Flaccid weakness Muscle wasting Muscle fasciculation Diaphragmatic and intercostals muscle weakness

# INTRODUCTION TO ALS

The ALS mortality rate is quite similar to the incidence rate due to short survival durations.

ALS knows no social, racial, or economic boundaries. Age specific incidence rates increase in men and women. The overall incidence and age specific incidence rates of sporadic ALS in men are more than those in women (1.6:1 to 1.3: 1). The incidence of ALS is rising, in part because of aging of the population into the high risk age groups, but the observed increase is not entirely due to demographic shifts.

## ETIOLOGY AND TREATMENT

To date, the etiology of ALS is unknown, although there are scientific theories of causation being researched around the world. Through understanding multiple interacting causes sharing a common pathway to motor neuron destruction, desperately needed therapeutic options will be developed. It is hypothesized a combination of therapeutic strategies to attack the disease at all levels will ultimately provide the means to alter the course of ALS.

People with ALS and their families may become intensely interested in potential causes of their disease. **It is important to emphasize that we don't have any reasonable evidence that could prove causation in an individual case.** Patients and loved ones need to be reassured that no lifestyle choices or past activity or exposure can be linked to their illness, that ALS is not contagious, and that in the absence of a family history, is not passed on to their children

Potential factors in mechanisms that might play a role in ALS and their relevance to the development of therapies are currently as follows:

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

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

- **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. In less than 10% of people with ALS, the disease is inherited. Seven genetic loci have been implicated in familial forms of ALS, each producing different features of the disease in the families affected. Every newly identified and located ALS gene provides scientists a piece of the ALS puzzle and creates the opportunity to develop new mouse models and cell lines that simulate these genetic abnormalities to research mechanisms that may occur in ALS.
- **Free Radicals and Oxidative Stress:** Canadian researchers have documented the existence of excessive levels of proteins damaged by free oxygen radicals within neurons in ALS, suggesting that either the neuron synthesizes excessive levels of free radicals, or that it is incapable of "venting" those that are normally produced. Therapies geared to reducing oxidative stress are in development, including gene therapy and new pharmacotherapy.
- **Immunological Factors:** In recent years, researchers have considered how injured motor neurons might spur an immune response that could contribute to, and even perpetuate, a cascade of cell death in the nervous system. The immune cells called microglial cells and astrocytes can respond to neural injury in a way that can either be beneficial or harmful. This microglial response has been implicated as a trigger of programmed cell death (PCD), a mechanism that is useful on a small scale to clear away damage, but devastating on a large scale as it ripples through the nervous system killing motor neurons.

# INTRODUCTION TO ALS

- **Neurotrophic Factors:** Although it is not clear how deficiencies of neurotrophic factors may affect human motor neurons, several attempts have been made to determine whether neurotrophic factors can slow the rate of progression of ALS by first testing these agents in animal models of the disease. Researchers are also investigating how the neurotrophic factors including brain derived neurotrophic factor (BDNF) and cytokine ciliary neurotrophic factor (CNTF) interact with metal ions in the cell, and how metal ions can thereby have extremely toxic effects in a cell depending on the neurotrophic factors present. Gene therapies are also in development to promote the levels of beneficial neurotrophic factors. The gene for insulin-like growth factor 1 (IGF-1) was successfully delivered in ALS mice using a viral vector, resulting in prolonging survival.
- **Altered Protein and Neurofilament Metabolism:** A signature feature of ALS is the accumulation of neurofilaments in the motor neurons. These key neuronal proteins are believed to be responsible for maintaining the normal neuronal structure and shape. Studies making use of transgenic mouse models to alter neurofilament expression reveal that abnormalities in the metabolism of neurofilaments, or the way in which neurofilaments interact with each other or with other proteins, could play a role in the development of ALS.
- **Glutamate Excitotoxicity:** Abnormalities in the handling of excitatory amino acids by the nervous system, particularly glutamate, may be critical to the occurrence of ALS. Through damage to the normal "transporter" mechanisms by which glutamate is removed from the nervous system, excessive glutamate accumulates. When motor neurons receive glutamate at their receptors, there is an influx in calcium ions into the cell. The motor neurons may not be able to deal with the excessive levels of calcium flooding in, resulting in damage. Researchers are investigating ways to help the nervous system handle calcium and glutamate. Riluzole, the single drug currently available for the treatment of ALS, shows very modest results. Its action is not well understood but is thought to perhaps affect glutamate mechanisms.

## ALS RESEARCH UPDATES

Ongoing basic scientific research into the cause and cure of ALS as well as clinical trials and studies are being conducted worldwide. For up to date information on research conducted in Canada and to download the Research Update Bulletins and the publication Northern Neuron, please visit [www.als.ca](http://www.als.ca). Additional scientific and clinical research information may be obtained by visiting [www.alsa.org](http://www.alsa.org), [www.wfnals.org](http://www.wfnals.org), and <http://clinicaltrials.gov>. For information on ALS clinical trials in Canada, visit the web site of the Canadian ALS Research Network (CALS), [www.alsnetwork.ca](http://www.alsnetwork.ca).

# PSYCHOLOGICAL SUPPORT

People with ALS, their families and caregivers (most often a spouse) often suffer considerable psychological and emotional distress. Much can be done to alleviate this distress, help people to adjust, and make the most of their coping skills. Treating someone with ALS requires great sensitivity, compassion and open-communication. When the need for specialized psychological support is identified, contact the ALS clinic team, social worker or nurse for an appropriate referral. Refer patients and families to the provincial ALS Society to learn about support groups and other available psychosocial support programs.

## BEFORE DIAGNOSIS

Recognize that patients and family will likely:

- Worry about symptoms
- Distress over difficulty in identifying the cause
- Be anxious due to required protracted period of investigation
- Express anger at perceived delays/uncertainties

## CONVEYING OR CONFIRMING DIAGNOSIS

Whether you are giving a possible diagnosis prior to confirmation by a neurologist, or seeing the patient for the first time after receiving their diagnosis from the neurologist (and in many cases due to stress they may not remember everything that was said) the following suggestions will apply:

- **Preparation** – take time to convey the potential seriousness of diagnosis and prognosis
- **Time and place** – the diagnosis should be discussed in a quiet, relaxed, private place away from external distractions; the patient's closest support system (spouse, partner, other family members) should be invited to be present
- **Amount of information** – initial shock often limits the ability to absorb information—let the patient know there is much to learn and there are resources available, but let them set their own pace for learning and respond accordingly; avoid absolute statements such as "ALS is fatal within 2 years" or "Unfortunately, there is nothing to be done."
- **Telling the truth** – honesty is important, but avoid leaving the person feeling alone and unsupported; prolonging uncertainty can exacerbate fear of the unknown; support the opportunity to come to terms with mortality and to make important decisions; identify immediate concerns and a plan to deal with them

## PATIENT'S EMOTIONAL RESPONSES

Common responses after an ALS diagnosis include:

- **Fear** – of increasing dependency and becoming a burden, also fearing the unknown, death and the process of dying
- **Denial** – is a coping mechanism that sometimes operates alongside awareness of the condition and its implications; professionals should accept this unless it is inhibiting appropriate support or palliation
- **Depression** – treatment of depression is likely to have a positive affect on ability to cope, but is not always easy to diagnose or differentiate from sadness and recognition that many of life's expectations can no longer be realized
- **Acceptance/adjustment** – coming to terms with reality is consistently challenged by proliferating impairments and increasing severity of symptoms—even an anticipated loss can impose new distress

## FAMILIES AND CAREGIVERS

- Consider strategies to balance the needs of the patient and other family members; caregivers who are coping well deliver better care
- Counteract individual isolation and promote awareness of each other's needs
- Create opportunities for caregivers to express negative feelings without guilt
- Identify resources for caregivers and family to better support both their need and those of the ALS patient

## IMPACT ON PROFESSIONALS

ALS frequently arouses strong emotional and ethical challenges:

- Attitudes toward issues such as disability, perceived quality of life, and measures taken to prolong life or hasten death may give rise to conflict over care decisions
- Frustration with the seeming inability to "fix" the problem can occur

Maintaining communication with other care team members can help you feel less isolated and helpless when treating an ALS patient.



# THE ALS HEALTHCARE TEAM

**A**ccess to healthcare professionals with expert knowledge of and experience with ALS who collaborate with the patient, family and primary care providers as a team is a very important factor in effectively managing ALS. There are several ALS specialty clinics in Canada that care for patients through an interdisciplinary approach. For a listing [Click Here](#). Your patient may have received their diagnosis from and be monitored by one of these teams. It is vital to your patient's quality of care and life that you communicate with the experts on the team to enhance your knowledge about ALS and appropriate symptom management.

## GENERAL ALS MANAGEMENT GOALS

- Provide information about the disease process, treatment options, and stay up-to-date with best practices
- Promote functionality and independence
- Communicate effectively with all other healthcare providers involved in the care plan to ensure seamless, well co-ordinated care
- Give people with ALS emotional support, hope, encouragement, and attention (time) in an accepting environment
- Respect the treatment preferences and priorities of people with ALS
- Involve family caregivers – provide and refer support as necessary
- Make ongoing assessments to guide timely referrals
- Suggest ways to manage the activities of daily living in a safe, efficient, and comfortable manner
- Assist in selecting, acquiring, and fitting assistive devices
- Liaise with community agencies that offer support and assistance accessing necessary equipment, funding and services
- Be open to discussions about death and dying, including reviewing advance directives and end-of-life care plans

## GENERAL NEEDS OF A PERSON WITH ALS

- Techniques and interventions for managing daily symptoms
- Ongoing teaching and counselling
- Straightforward information about the disease process
- Access to equipment and services to promote functionality and quality of life

- Timely evaluations of the progression of the disease
- Anticipation of future needs and care planning

## HEALTHCARE TEAM ROLES

While all teams may not have all of the following disciplines represented, the following list includes the make-up of an ideal interdisciplinary care team for best possible care and support.:

### Primary Care/Family Physician

- Explains the diagnosis and possible progression of the disease
- Provides ongoing management of pre-existing conditions
- Continues general medical care including primary prevention measures (e.g., flu shots)
- Treats symptoms
- Makes necessary referrals to, and consults with, other healthcare providers and community agencies to best manage care
- Discusses end-of-life issues including advanced care directives
- Provides support and encouragement to the patient and family
- Assesses the patient regularly to maintain continuity of care

## NEUROLOGIST (WITH ALS SPECIALTY)

- Confirms ALS diagnosis
- Outlines types of treatment options available
- Encourages the setting of short-term goals
- Helps the patient preserve a positive self-image and maintain morale
- Works with patient to identify specific needs and concerns and refers to therapists who may be able to find solutions
- Collaborates with primary care physician and all other caregivers about care
- Sees patient for follow-up every 3-4 months

## ALS NURSE (NEUROMUSCULAR SPECIALTY)

- Develops a care plan
- Explains terminology and techniques
- Teaches skills and provides demonstrations

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# THE ALS HEALTHCARE TEAM

- Addresses questions and concerns
- Limits the development of complications
- Ensures comfort
- Facilitates decision-making by the people with ALS and their families
- Collaborates with other team members and community agencies
- Provides information and support to families and care-providers unable to attend clinic

## OCCUPATIONAL THERAPIST

- Assesses fine-motor functional abilities
- Evaluates positioning and seating requirements
- Assists in choosing and accessing suitable assistive devices and strategies for maintaining independent function
- Gives 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
- Teaches energy conservation and time management techniques
- Makes home assessments and provides information about modifications to home and other environments to enhance mobility and safety

## PHYSIOTHERAPIST

- Provides a detailed analysis of abnormal movement
- Optimizes patient strength, function, and comfort
- Designs and monitors therapeutic exercise regimen when appropriate
- Assists with breathing management/airway clearance
- Provides training in energy conservation and time management techniques
- Provides advice on suitable equipment to maximize mobility
- Makes home assessments

## PHYSIATRIST (PHYSICAL AND REHABILITATIVE MEDICINE PHYSICIAN)

- Recommends supportive treatment
- Prevents complications through timely treatments
- Determines which diagnostic tests are necessary
- Helps maintain maximum function and quality of life
- Consults with therapists concerning orthoses and assistive equipment

## DIETITIAN

- Assesses functional abilities related to eating
- Recommends appropriate changes in food texture and consistency
- Suggests methods for food preparation
- Advises substitutions for hard-to-manage foods
- Provides strategies for maximizing nutritional intake and hydration

## GASTROENTEROLOGIST

- Oversees nutritional care of patients
- Arranges for feeding tubes

## RESPIRATORY THERAPIST AND RESPIROLOGIST

- Evaluates pulmonary function status
- Devises strategies to optimize remaining breathing function and reduce discomfort
- Offers information on body positioning, energy conservation, relaxation, and compensatory techniques to improve breath support for nutrition and speech
- Teaches caregivers how to operate non-invasive ventilators
- Makes suggestions about a course of action when respiratory failure occurs
- Treats respiratory infections

## SPEECH-LANGUAGE PATHOLOGIST (SLP)

- Evaluates functional abilities, such as oral motor function, cognitive-linguistic function, augmentative communication function, and swallowing function
- Evaluates the individual's motivation and potential for learning new techniques
- Determines the most efficient communication function
- Recommends appropriate technology for augmentative communication
- Trains people with ALS and family members in techniques of effective communication and energy conservation

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# THE ALS HEALTHCARE TEAM

## SOCIAL WORKER

- Offers emotional support and counselling to the person with ALS and to family members
- Provides information about available community resources and acts as an advocate to assist people in accessing these resources
- Provides information on legal and financial issues and assists people to access these resources in their communities

## PALLIATIVE CARE PHYSICIAN

- Assists patients with decision-making throughout the disease process about various care options including respiratory or nutritional interventions, preferred place of care, personal goals
- Discusses end-of-life issues, concerns, fears, wishes
- Consults on pain management and other care aimed at improving comfort and quality of life for patients

## PASTORAL CARE WORKER

- Listens to and empathizes with those who want to vent concerns
- Assists in decision making
- Gives spiritual support during emotional or physical crises
- Provides liaison to community pastoral and spiritual services
- Acts as an advocate for those who have no voice
- Encourages people with ALS to discover their personal strengths
- Encourages people with ALS to recapture positive experiences from the past by recall or reminiscence
- Listens to concerns about death and dying
- Arranges and conducts services when appropriate

Healthcare providers are advised to form an ongoing partnership with ALS patients and their families to devise and maintain a care plan oriented to the person, not the disease.

The ALS team including healthcare professionals, families, friends and the ALS Society, plays an important role in educating homecare providers who may be unfamiliar with the unique needs of ALS clients.

# DYSARTHRIA

**D**ysarthria seen in ALS is the mixed flaccid-spastic type. Speech is affected by weakness, slowness and sometimes paralysis of the muscles of the lips, tongue, jaw, soft palate and larynx. Breathing changes can also result in voice changes as well. In severe cases, anarthria results with complete loss of intelligible speech. Sensation is usually intact.

## CLINICAL FEATURES

Impairment of speech production (dysarthria) may begin with:

- Slurring especially with fatigue
- Hoarseness
- Breathy voice

Patient's emotional response can be:

- **Isolation** – communication inadequate or avoided
- **Frustration** – difficult or impossible to be understood, need time which may not be available
- **Increased fear and anxiety** – because patients are unconventional communicators, others assume hearing or intellectual impairment which increases anxiety around failed communication attempts
- **Loss of control** – because misunderstood or opinion ignored or not solicited
- **Increased sadness** – isolation and frustration felt by patient, care person and family

## MANAGEMENT

Early referral to **Speech Language Pathology** for:

- Advice on strategies for speech clarification strategies, augmentative and alternative communication (AAC)
- Assessment for and provision of communication aids and training in their use
- Education and support of patient, family, and other communication advocates

The **speech language pathologist** will work with an **occupational therapist** who can advise on positioning, wrist supports, switches, pointers, mobile arm supports and tables, and other access methods for communication devices.

## CONSIDERATIONS TO ENHANCE PROVIDER-PATIENT COMMUNICATION

- Encourage patient to slow down and carefully exaggerate articulation of sounds
- If there is no intelligible speech, establish reliable easily reproducible gestures or signals for yes and no; ask questions that only require a “yes” or “no” answer
- Position face-to-face: watch lips, eyes, gestures
- Take time to create a relaxed, quiet environment for communication
- Avoid interrupting or trying to finish sentences. Ask if they wish you to complete sentences or guess at words – some appreciate the saved effort, others dislike the interruption.

## COMMUNICATION CAN BE AIDED BY:

- Writing if hand function is good
- Alphabet board
- Perspex eye pointing frame (e.g., ETRAN BOARD)
- Hands-free telephone
- Call bells
- Personal alarms
- Computerized communication aids, (e.g., hand held LIGHTWRITER™)
- Telephone typewriter (TTY)
- Communication computer software

*NOTE:* The provincial ALS Society and therapists working with the patient are excellent resources for ACC information, equipment, and programs.



# DYSPHAGIA AND NUTRITION

**D**ysphagia, including chewing and swallowing problems, is caused by weakness of the lips, tongue, masseter muscles, soft palate and esophagus. Oral, pharyngeal, and esophageal phases of swallowing, or a combination, may be affected. Often, the first symptoms include coughing on thin liquids and dry, crumbly textures.

**Nutrition** is an independent prognostic factor for survival in patients with ALS. The disease and its progression (i.e., dysphagia, fatigue, pulmonary involvement) affect nutritional intake. Nutritional status can affect muscle function, respiratory function and quality of life. There is no special diet for ALS – the recommendation remains a nutritionally well-balanced diet.

## CLINICAL FEATURES OF DYSPHAGIA

- Impaired ability to chew, form a bolus and move it posteriorly with the tongue
- Delayed or effortful swallow onset
- Reduced or inconsistent airway protection during the swallow

Dysphagia can lead to drooling, muscle spasms, malnutrition, dehydration and aspiration with increased risk of aspiration pneumonia.

## MANAGEMENT

- Rapid, co-ordinated, interdisciplinary healthcare team approach
  - ▶ Early referral to **speech language pathologist** for a swallowing assessment and recommendations for safe swallowing
  - ▶ Referral to **registered dietitian** for additional nutritional strategies such as food and liquid texture modification, foods to avoid, supplements, and behavioural strategies, i.e., eat without distractions
  - ▶ Referral to **occupational therapist** for advice on adapted feeding tools, modified plates and cups, non-slip mats, arm supports etc.
- Ongoing assessment and management of changes in swallowing function is required to minimize the risk of malnutrition, dehydration, and aspiration
- These services are usually available through the ALS clinic



## NUTRITIONAL IMPLICATIONS


- There is a **hypermetabolic nature of ALS**—weight loss in ALS results from loss of muscle mass, decreased intake as well as from energy cost of activities
  - ▶ Discuss maximizing caloric intake and energy conservation strategies
- Assess bowel function for constipation resulting from delayed transit time or decrease in activity, inadequate fibre intake, low fluid intake, and medications such as opioid analgesics and anticholinergics
  - ▶ Constipation management includes adequate hydration, adequate dietary fibre, regular meal times, establishing a bowel routine, medication management, stool softeners and, if necessary, laxatives
- Monitor weight loss closely, discuss placement of feeding tube, and help patient in decision-making process
- Consult with ALS clinic for referral to a gastroenterologist for tube insertion

## TUBE FEEDING

Oral feeding may become a high risk activity due to increased risk of obstruction of the airway, aspiration and inability to meet nutritional requirements.

**Percutaneous endoscopic gastrostomy (PEG)** has become the intervention of choice when eating becomes exhausting, difficult or nutritional goals are not being met because oral intake is time-consuming and onerous. PEG placement does not eliminate the risk of aspiration. In some ALS centres the tube is inserted into the jejunum as opposed to the stomach—this is called a "J-tube."

Having a tube inserted does not preclude oral feeding as well when appropriate. While there are obvious benefits to having a feeding tube, the decision to have one remains the patient's. A useful educational tool for discussing with your patient the advantages and disadvantages of having a PEG is found using these links  

**NOTE:** Timing of PEG is dependant on breathing function. Consult with your patient's neurologist and the respirologist on the ALS treatment team. 

# DYSPNEA

**S**hortness of breath (dyspnea) is a common symptom in the course of ALS. Usually respiratory muscle weakness occurs late in the disease and contributes to the most likely cause of death, respiratory failure. However, ventilatory failure can develop at any stage and occasionally is the presenting feature of ALS. **Shortness of breath may not be experienced or reported as a breathing symptom; fatigue, anxiety, claustrophobia, and insomnia may all represent reduced respiratory function.**

## CLINICAL FEATURES

- Breathlessness on exertion, sometimes just the exertion of speaking or eating
- Sleep disturbance, anxiety, and panic
- Orthopnoea—breathless lying flat (most patients find a semi-recumbent position the most comfortable; this position allows the intercostal muscles and the diaphragm to work to their greatest mechanical advantage and is more comfortable for swallowing)
- Hypoventilation is worse during sleep (upper airway may also be partially obstructed because of bulbar and laryngeal muscle weakness)
- Increasing CO<sub>2</sub> levels in the blood may result in headaches, nausea, somnolence, and encephalopathy, especially on waking
- Hypoxemia if there is co-existing lung disease

## MANAGEMENT

- Discussion of respiratory management must be initiated well in advance of first symptoms (unless present at the time of diagnosis)
- Choice of respiratory interventions must be clearly presented:
  - ▶ Medical only
  - ▶ Non-invasive ventilation (NIV, often BiPAP™)
  - ▶ Tracheostomy and long-term invasive mechanical ventilations (IMV)
- Assurance that the best medical management will be provided both with NIV or IMV, and should withdrawal be requested for either
- Anxiety relief – reassurance and/or medication
- Controlled breathing exercises and assisted coughing techniques taught by respiratory therapist or physiotherapist

- Correct posture in chair and bed (beds with adjustable back supports and recliner chairs may be helpful)
- Secretion control (retained secretions in the mouth and pharynx further compromise the airway and add to the patient's discomfort and panic)
- Suction apparatus may be of benefit

**NOTE:** Oxygen therapy in ALS:

- ▶ Oxygen therapy in the setting of CO<sub>2</sub> retention may lead to increasing hypercarbia, coma, and death, therefore oxygen should be used with caution. Oxygen can dry nasal membranes. The apparatus may be noisy, making communication more difficult. The mask or prongs may be uncomfortable.
- ▶ Airflow on the V2 territory of the fifth cranial nerve has been shown to provide comfort – this could be in the form of a fan, or room air via mask or prongs
- ▶ However, if the patient finds relief of symptoms with oxygen rather than airflow alone, then oxygen is appropriate for palliation
- ▶ Clarify treatment goals before any recommendations for oxygen therapy are made

## CONSIDER

- Regular monitoring of pulmonary function: vital capacity, cough ability
  - ▶ This can provide the opportunity to discuss respiratory issues as well as anticipate respiratory decline
- Measuring arterial blood gases, pO<sub>2</sub>, and pCO<sub>2</sub> (this gives some guidance to the prognosis and also indicates whether oxygen therapy is appropriate)
- Referral to a respirologist if some form of assisted ventilation is appropriate and is desired by the patient
  - ▶ NIV – either intermittent non-invasive positive pressure ventilation (NIPPV) or bi-level positive airway pressure device (BiPAP™) by mask has been shown to improve quality of life, and may meaningfully prolong life
  - ▶ IMV if patient's goal is long-term survival (and appropriate supports are in place) rather than delay of dying

# DYSPNEA

## IMPLICATIONS OF MECHANICAL VENTILATION

- Quality of life issues must be addressed
- Increased dependency on family and caregivers, especially in case of IMV
- Increased need for professional nursing support—is appropriate nursing care available and affordable in the case of IMV at home?
- Death occurs due to voluntary or accidental discontinuation of IMV, from complications of IMV or from a new condition, not due to ALS directly
- Advanced medical directives – at which point should ventilatory support be withdrawn? Do the patient and family have personal and spiritual support to withdraw therapy if started?


## PALLIATION


Breathlessness is one of the most frightening symptoms of ALS. Few patients plan to use invasive mechanical ventilation but **the decision to choose invasive ventilation support should not wait until respiratory symptoms have developed.** For those who can tolerate NIV, symptoms of CO<sub>2</sub> retention and dyspnea may be alleviated for months and, rarely, years. Bulbar weakness may limit NIV use, and as well as the progression of respiratory muscle weakness. If invasive ventilation is not chosen, or it is being discontinued, recommendations for relieving dyspnea and anxiety are as follows:

- Palliation of dyspnea is best achieved using **opiates and in consultation with a palliative care team and/or specialist ALS care team:**
  - ▶ Incremental increases in opioids (e.g., morphine or hydromorphone) are often required to control symptoms of dyspnea. Usually patients benefit from a bedtime dose, but may use it intermittently in the daytime. Duration of action is a maximum of four hours. Gradually increase dosing and frequency as required.
  - ▶ When anxious or distressed by breathlessness or retained secretions in the throat, a small dose 4 to 5 minutes before meals can improve symptoms of dyspnea while eating
  - ▶ More regular doses of morphine may be required in the more terminal stages of the disease for control of symptoms. The use of slow

release morphine preparations or subcutaneous (s/c) delivery via a syringe driver may need to be considered in these late stages, particularly if the patient chooses to die at home

- ▶ Another approach is to order subcutaneous morphine or dilaudid and then switch to fentanyl patches if the doses are equivalent  
**(Important: Contact pharmacist for assistance in opioid conversions)**

*NOTE:* Morphine can be delivered orally or subcutaneously, but in equivalent doses, s/c is 2-4 times more potent. 

*NOTE:* Predictably swallowing a long acting preparation of opioids needs to be determined before prescribing—contins cannot be given through a g-tube because they cannot be crushed. 

- Other drugs:
  - ▶ Benzodiazepines – small doses of diazepam or clonazepam may be added to help control anxiety
  - ▶ Anticholinergics – hyoscine, atropine, amitriptyline, glycopyrrolate reduce secretions in the airways of dying patients

Stress the message to patients that **deaths caused by choking attacks are almost unheard of in ALS** and that the final stages of the disease are usually peaceful and dignified.

# SIALORRHEA

**S**ialorrhea, or **drooling**, occurs not because of excessive saliva production but rather a decreased ability to manage normal saliva production due to impaired control of tongue, lips, and swallowing muscle function. A build-up of saliva and thick mucous can disrupt sleep and increase the risk for choking.

*NOTE:* When treating with medication, remember to consider side effects that may increase risk of falling due to drowsiness or slowing breathing.



*NOTE:* Even edentulous patients should have regular inspections of the intra-oral cavity.



## CLINICAL FEATURES

- Anterior pooling of secretions resulting in less triggering of swallow to clear saliva
- Poor lip seal, that would otherwise prevent leakage, can produce the perception that there is excessive saliva production

## MANAGEMENT

- May include anticholinergic medications including atropine and amitriptyline, or scopolamine patches (side effects such as excessive mouth dryness and constipation are reported)
- Individual patients may report improvement with decreasing the amount of dairy products consumed
- Conscientious and regular oral hygiene is important for all patients (including those receiving all nutrition and fluids via feeding tubes) to maintain oral-dental health; reduce harmful intra-oral bacteria production; and reduce the risk of pneumonia from aspiration of potentially "unhealthy" secretions
- Portable suction device may be useful in some instances
- In some ALS centres, botulinum toxin injections into the parotid (by a trained neurologist) or irradiation of the salivary glands may be offered to patients with medically refractory sialorrhea
- Rarely, choking on thick secretions is sufficiently life-threatening that tracheostomy for secretion management may be the only way to palliate this symptom. This should be offered even to those who do not want long-term invasive mechanical ventilation – the decision to use tracheostomy in this situation is to prevent suffering caused by airway obstruction.



# PSEUDOBULBAR AFFECT

**P**seudobulbar affect, or **emotional lability**, is characterized by uncontrolled and inappropriate crying or laughing. It is thought to occur as a result of lost inhibition of the limbic motor neurons in the brain which control muscles involved in primitive vocalization. Emotional lability is **not a mood disorder, but abnormal affective display**. Physicians must be alert to pseudobulbar affect as it is often not recognized as part of ALS: it can be confused with depression; and patients often do not report symptoms, yet as many as 50% of ALS patients experience pseudobulbar affect.

- ◆ Valproate
- ◆ Lithium

***NOTE:*** An investigational drug, **Neurodex™**, a combination of dextromethorphan and quinidine, has shown efficacy in clinical trials for the treatment of emotional lability in ALS patients. An open-label study to assess long-term safety is being conducted in the U.S. Although not yet available, it may be a treatment to watch for in the future.

## CLINICAL FEATURES

- Episodes of uncontrollable laughing or crying which may be inappropriate or unrelated to the situation at hand
- Crying may be prolonged and difficult to stop
- Expression may be spontaneous or unmotivated
- Discussing topics with emotional content may trigger laughter or crying that is disproportionate, (e.g., speaking about a child's graduation, which would have an emotional connection, may trigger sobbing)
- Most common in ALS patients with difficulty swallowing and speaking
- Excessive uncontrolled crying more common than laughing

***NOTE:*** Pseudobulbar affect can have significant impact on relationships, social interaction, and overall quality of life. If not effectively managed, high levels of associated frustration and social anxiety can result in social withdrawal.

## MANAGEMENT

- A proper diagnosis distinct from depression is critical as treatment approaches using antidepressants will differ (may require a neurological consult; assessment tool for pseudobulbar affect is also available to physicians)
- Recognition and discussion of the social management of the symptom may be sufficient, but if intervention is required,
  - ▶ Consider the following antidepressants:
    - ◆ Tricyclic antidepressants
    - ◆ Selective serotonin reuptake inhibitors

# PAIN

**P**ain and discomfort in ALS arise as complications of muscle weakness, stiffness, and immobility.

## CLINICAL FEATURES

- Loss of muscular control to stabilize large joints and maintain spinal posture
- Passive injury to joints when controlling muscles are weak, (e.g., shoulder joint damage during assisted transfers)
- Neuropathic pain from entrapment or positioning (e.g., foot drop, and some patients complain of burning, or pins and needles in their hands and feet)
- Exacerbation of previous pain syndromes, back pain, etc.
- Muscle cramps
- Spasticity (including jaw spasms)
- Skin pressure
- Constipation
- Laryngospasm

## PAIN MANAGEMENT

### Positioning and Physiotherapy

Co-ordination with a physiotherapist is essential to determine the relative role of medication and regular physiotherapy in managing pain.

- Early active stretching programs while muscle strength is retained may prevent joint and muscle pain developing
- Later in the course of the disease, careful positioning in bed and chair to support head, trunk and weight of limbs
- Regular repositioning for patients unable to reposition themselves
- Passive limb movements to prevent muscle and joint stiffness
- Physiotherapy advice on most appropriate passive exercise, positioning and transferring techniques, and pressure relieving equipment (e.g., special cushions, mattresses or pads)

### Drug Therapy

#### 1. Analgesia:

- Non-steroidal anti-inflammatory drugs if there is an

arthritic or inflammatory component to the pain

- Intra-articular injection, especially into the shoulder joint
- Small doses of oral morphine are often very helpful
  - ▶ Start with very low doses (e.g., 2-4mg at night and increase gradually if necessary). The dose may be repeated in 4-hour increments.
  - ▶ Consider slow-release morphine preparations if regular analgesia is required
  - ▶ Always institute a bowel regimen if not already in place
- Anticonvulsants – gabapentin has the lowest side-effect profile, but may be a cost issue
- Antidepressants – tricyclics such as amitriptyline and desipramine have anticholinergic side effects, which may dry secretions which could be helpful in ALS patients who have neuropathic pain and excessive salivation

#### 2. Anti-Cramping/Spasm Drugs:

*NOTE:* Cramping more common early in the disease course. 

- Quinine bisulphate ~300mg at night
- Diazepam
  - ▶ Consider reviewing the use of diazepam as its duration of action is so long and becomes longer with increasing age. Although, the long duration of action may be its benefit in patients who tolerate it well.
- Baclofen (side effects of drowsiness, muscle weakness and rash may occur)
  - ▶ For cramping start with very small doses, (e.g., baclofen 5-10mg bid, or diazepam 1-2mg). Only bedtime doses may be required.
  - ▶ For spasms start with 5-10mg bid. Gradually increase dose as required. It is seldom worth exceeding a total daily dose of 75mg.

*NOTE:* Your Provincial ALS Society may have pressure relieving equipment available to ALS patients through its Equipment Program. Encourage your patient with ALS to register with the Society to benefit from this and other programs and services.

# PALLIATIVE CARE

**A**LS is presently incurable and there is no effective treatment, thus, **palliation is part of the continuum of care from diagnosis onward.**

In ALS, symptom control and promoting quality of life throughout the disease process is considered palliative care—it is not limited only to hospice, or comfort care, most commonly associated with end-stage cancer care. Palliative care focuses on the person, not the disease, to help them achieve how they would like to live out their life in the time remaining.

Another aspect of care unique to a disease such as ALS is that your patients will experience a series of functional losses throughout the duration of the disease, each accompanied with issues of grief and bereavement. Be aware that loss and grief do not only pertain to dealing with death and dying for ALS patients and family.

**Goals of treatment should be discussed with patients and family and repeated as a patient's status changes.** As ALS progresses to a terminal phase, the goal of palliative care changes from maximizing function to providing compassionate hospice care to ensure a peaceful and dignified death according to the patient's beliefs and preferences.

## PALLIATIVE CARE IN PRACTICE

- **Issues unique to ALS:**
  - ▶ Understand the differences in palliative care in ALS vs cancer
  - ▶ Reassure the patient you will not abandon them even though you cannot offer them a probable cure; too often ALS patients report feeling abandoned after diagnosis
- **Referrals and links:**
  - ▶ Establish links at an early stage with palliative care organizations and professionals to obtain useful advice and support for working with a patient and/or caregiver who may need additional time to come to terms with the terminal nature of the disease
  - ▶ Refer to the palliative care team or palliative care specialist for guidance in providing comfort care and grief and bereavement counseling when needed

- ▶ Recognize cues from patient signaling a need or desire for spiritual counseling and make appropriate suggestions and referrals
- **Advance care planning:**
  - ▶ Keep lines of communication open regarding end-of-life issues to identify the right time to discuss advanced care planning and make appropriate referrals when necessary
  - ▶ Practice shared decision making based on unbiased professional explanations, personal preferences, and religious or cultural beliefs
  - ▶ Revisit with your patient, and their caregiver who will act as their proxy, their advanced care directives at least every 6 months (as choices may change during the course of the disease)
  - ▶ Keep all appropriate healthcare team members informed of your patient's current end-of-life care choices
- **Complementary care:**
  - ▶ Explore use of complementary therapies that have been found to help some patients: aromatherapy, reflexology, massage, relaxation techniques, and acupuncture

## CARE IN THE TERMINAL PHASE

For some people death can be very sudden, often before an obvious end-stage is reached. Others experience a protracted final stage which can last for many weeks. The most common cause of death in ALS is respiratory failure usually following respiratory tract infection.

*NOTE:* It is important to reassure patients and families that death from choking is rare.



### Action:

- Check all symptom management in the following areas:
  - ▶ Pain
  - ▶ Dyspnea
  - ▶ Dysphagia
  - ▶ Sialorrhea
  - ▶ Insomnia
  - ▶ Anxiety & depression
  - ▶ Bowel & bladder function

# PALLIATIVE CARE

- Maintain communication with the patient as long as possible:
  - ▶ Eye pointing or single response answers to closed-ended questions can be maintained
- Reassess emotional and practical needs of the family and caregiver such as sharing information about care plans, and facilitating adequate nursing coverage if the patient is at home

## Medication:

- Incremental increases in opioids (e.g., morphine or hydromorphone) are often required to relieve dyspnea and control pain (see Dyspnea section for more specific medication recommendations on end-stage palliation of dyspnea)
- Opioids are also used to reduce cough reflex
- Anti-cholinergics such as hyoscine hydrobromide reduce saliva and lung secretions, and relax smooth muscle
- Sedatives such as diazepam, midazolam, chlorpromazine reduce anxiety

**Good symptom control is essential in both the management of living with ALS as well as for a peaceful and dignified death.**

# EQUIPMENT TO MAINTAIN INDEPENDENCE

**A**ssistive 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 ongoing need for different assistive devices to maintain optimal functionality. Provincial ALS Societies provide assistance in obtaining equipment through their Equipment Programs when other avenues have been exhausted (public and private health benefits).

People should not purchase equipment or modify their home or vehicle unless they have discussed options with appropriate healthcare professionals. Equipment and renovations can be costly and take time to obtain. Each piece of equipment is different and has special features that may not be suitable for every person with ALS. Healthcare professionals such as **occupational therapists** familiar with available equipment can answer the following:

- Advantages and disadvantages of the equipment or renovation
- Safety issues that need to be considered
- Immediate and long-term needs
- Funding or loan options will help the person choose the solution that will best serve their needs

## ASSISTIVE TOOLS FOR ACTIVITIES OF DAILY LIVING (ADL)

Many people with ALS develop trouble holding objects. It is possible to modify everyday tools or to purchase aids to compensate for weakness in the muscles of the fingers, hands, and wrist:

- Utensils with extra thick handles
- Mugs with extra thick handles
- Thick pens or pencils
- Doorknob turner
- Velcro or elastics are a popular replacement for both buttons and zippers on clothing
- Hands-free phone
- Holder for playing cards
- Environmental controls that enable a person with ALS to turn on lights, radio, television, open and close doors, start a coffee maker, etc., with the palm of the hand, head movement, or even a puff of breath

## BODY SUPPORTS (MAJOR ORTHOSES)


Various types of body supports available through hospitals and clinics are used to support joints in certain positions when muscles weaken, and prevent contractures if spasticity is a problem. These items may increase comfort and prevent fatigue as well as improve a person's ability to do their activities of daily living:

- Ankle and foot orthoses (AFOs)
- Hand and wrist supports
- Shoulder (sling) and neck (cervical collar) supports

## MOBILITY

The ability to move around independently and safely is important to maintain a person's activity in their home and community. Conserving energy by using different equipment for mobility allows a person to participate in activities he or she enjoys. For example, a person may use a cane around the house but use a power wheelchair for longer distances. Precautions related to environmental safety must be implemented when the person with ALS is using mobility equipment (e.g., remove carpets that may increase risk for tripping, wheelchair getting caught). It is important not to wait until the person cannot move without assistance; talk with the therapist as soon as possible as it takes time to obtain equipment:

- Canes and walkers
- Manual wheelchairs
- Power wheelchairs
- Tilt and recline functions on wheelchairs
- Cushions chosen for comfort and pressure relief
- Scooters

**NOTE:** Getting a proper wheelchair seating assessment from a therapist who specializes in seating is vital for comfort, safety, and mobility. 

## LIFTS AND STAIR GLIDES

Various options for equipment to move a person to a different level such as from one floor of the house to another or from the tub to a wheelchair are available:

- **Chair Lift** – automated recliner chair which moves from sitting to standing position

# EQUIPMENT TO MAINTAIN INDEPENDENCE

- **Ceiling Lifts** – a small lift is attached to a track in the ceiling to transport a person from the bed to bath, wheelchair, toilet
- **Portable Lifts** – portable lifts such as Hoyer Lift use a sling to lift a person up. This type of lift is moveable throughout the house
- **Stair Glides** – an automated chair can be moved up and down a track to move from one floor to another
- **Wheelchair Lifts** – can take a person up or down a level inside or outside the home when other alternatives such as ramps are not appropriate

## BATHROOM EQUIPMENT

- Raised toilet seats or commode chairs
- Bidet attachments to toilet to maintain independence toileting
- Bathtub seats or lifts
- Superpoles to assist with a standing transfer

## AUGMENTATIVE AND ALTERNATIVE COMMUNICATION (AAC) EQUIPMENT


The loss of functional speech from ALS can be devastating and lead to isolation unless steps are taken early to provide other ways of communicating. **Education and assessment should be started long before speech therapy techniques or speech amplifiers cease to keep speech viable.** It can be encouraging to know that many AAC options exist, ranging from no-tech/low-tech (e.g., letter boards for spelling) to high-tech (e.g., sophisticated voice output communication aids – VOCAs), and many options in between.

More often than not, rather than a single device or communication method, a communication **system** is needed that is comprised of both low-tech strategies and one or more electronic devices. This is because no single communication method may be good for all types of communication in all environments, and needs can change over time (e.g., computers are not good in the bathtub, or someone who can write now may not be able to in a few weeks or months).

Once a comprehensive system has been appropriately prescribed, if private health benefits or provincial

equipment programs cannot fund or provide all the required technology, Provincial ALS Societies may be able to offer assistance in the procurement of technology.

Here are a few examples of the multitude of devices and techniques available:

- Clear plastic communication boards (low tech) which users with no arm function can still point to with their eyes to spell or select messages
- Erasable writing boards or slates for those who can still write but don't want to carry around a lot of paper
- Speaking valves for some who have mechanical ventilation through a breathing hole, but who still have good articulation for forming words
- Portable VOCAs, also known as SGDs (speech generating devices) that have a speech synthesizer that will speak anything spelled into them or can play pre-stored messages at the touch of a button
- VOCAs/SGDs created by using special communication software on a portable or desk-top computer 

**NOTE:** It is critical patients are assessed by an assistive technology practitioner, who in most provinces would be either a **speech language pathologist** or an **occupational therapist**, with special training and experience in prescribing AAC and in teaching users how to operate their systems. Proper assessment and prescription will prevent expensive, inappropriate purchases while expert training will facilitate optimal use of a device.

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# RECOMMENDED READING AND OTHER RESOURCES

This list is meant to introduce you to some texts and resources that will help you as a professional better understand ALS and the needs of the ALS patient. A more comprehensive list of resources to recommend to your patients is found in the *Manual for People Living with ALS*. Check with your patient to see if they have this manual. If not, direct them to their provincial ALS Society to obtain one or to download it from [www.als.ca](http://www.als.ca).

## MEDICAL TEXTS ABOUT ALS:

### **Amyotrophic Lateral Sclerosis**

By Hiroshi Mitsumoto, MD, 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

### **Completing the Continuum of ALS Care: A Consensus Report**

By Hiroshi Mitsumoto, MD, DMedSc, ALS Peer Workgroup Chair, and The ALS Peer Workgroup Members. This publication was produced by Promoting Excellence in End-of-Life Care, a national program of The Robert Wood Johnson Foundation, directed by Ira Byock, MD.  
[www.promotingexcellence.org/als/index.htm](http://www.promotingexcellence.org/als/index.htm)

### **Motor Neuron Disorders**

Edited by: Pamela J. Shaw, MD and Michael J. Strong, MD  
Butterworth-Heinemann/Elsevier, Inc.  
Independence Square West  
Philadelphia, PA 19106  
(215) 238-2239 ISBN: 0750674423

### **Palliative Care in Amyotrophic Lateral Sclerosis: Motor Neuron Disease**

By David Oliver, MD, Gian Domenico Borasio, MD, and Declan Walsh, MD  
Oxford University Press  
ISBN: 0192637667

## ALS MANAGEMENT GUIDES AND COPING WITH CHRONIC ILLNESS:

### **A Manual for People Living with ALS**

Produced by the ALS Society of Canada  
Editor, Jane McCarthy, MSc, MPH  
This comprehensive manual covers background information about the disease ALS, theories of causation, symptom management, coping with ALS, end-of-life issues, financial and legal issues, and an extensive, descriptive list of educational and informational resources, and record keeping and communication tools. It can be downloaded or a hardcopy can be obtained by patients and family members, through provincial ALS Society offices at no charge. All other orders for a hardcopy are subject to a fee.

[www.als.ca](http://www.als.ca)

1-800-267-4257 - 1-905-248-2052 (Greater Toronto Area)

### **Amyotrophic Lateral Sclerosis**

By Robert Miller, MD, Deborah Gelinas, MD, and Patricia O'Connor, RN  
This is one of the first in a series sponsored by the American Academy of Neurology designed to assist people living neurologic diseases and their families. This book is an excellent resource for those confronted with newly diagnosed ALS. It includes chapters about the disease itself, symptoms and how they can be lessened, how multidisciplinary ALS clinical centres work, how voluntary health agencies help, how computers can help, how the internet can be used, and how to deal with health insurance.

AAN Press

Demos Medical Publishing, Inc.  
386 Park Avenue South  
New York, NY 10016 ISBN: 1932603069  
[www.demosmedpub.com](http://www.demosmedpub.com)



# RECOMMENDED READING AND OTHER RESOURCES

## **Amyotrophic Lateral Sclerosis:**

### **A Guide For Patients and Families, 3rd Edition**

By Hiroshi Mitsumoto, MD & Theodore L, Munstat, MD

This comprehensive guide covers every aspect of the management of ALS. Beginning with discussions of its clinical features of the disease, diagnosis, and an overview of symptom management, major sections deal with medical and rehabilitative management, living with ALS, managing advanced disease and end-of-life issues, and resources that can provide support and assistance.

Demos Medical Publishing, Inc.

386 Park Avenue South, New York, NY 10016

ISBN:1888799285

[www.demosmedpub.com](http://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

### **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 her parents, one with cancer and the other ALS. Through reading this book, healthcare professionals can gain special insight into the needs of ALS family caregivers.

Wiley and Sons Publishing

ISBN: 0471254088

### **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

### **Learning to Fall: The Blessings of an Imperfect Life**

By Philip E. Simmons, PhD

Read an excerpt at [www.learningtofall.com](http://www.learningtofall.com),

Xlibris: [www.xlibris.com](http://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

### **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)**

### **Motor Neuron Disease: A Family Affair**

By: David Oliver, MD

Sheldon Press

36 Causston St, London, SW1P 4ST

ISBN: 0859699773

### **What If It's Not Alzheimer's? A Caregiver's Guide to Dementia**

Edited by Lisa Radin and Gary Radin

Foreword by Murray Grossman, MD, EdD

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

# RECOMMENDED READING AND OTHER RESOURCES

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

## COOK BOOKS:

### **The Dysphagia Cookbook: Great Tasting and Nutritious Recipes for People with Swallowing Difficulties**

By Elaine Achilles, EdD  
Cumberland House Publishing  
431 Harding Industrial Drive  
Nashville, TN 37211  
ISBN: 1581823487

## VIDEOS:

### **The ALS Association's Living with ALS Video Series:**

1. Clinical Care Management Discussion with ALS Experts
  2. Mobility, Activities of Daily Living, and Home Adaptations
  3. Adapting to Breathing Changes and Non-Invasive Ventilation
  4. Adjusting to Swallowing Difficulties and
  5. Maintaining Good Nutrition
  6. 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](http://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](http://www.films.com)

### **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.

<http://www.lm-media.com/videos/>

### **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](http://www.lesturnerals.org)).

## INTERNET RESOURCES:

### **Other ALS Organizations:**

- [www.alsmndalliance.org](http://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](http://www.alsa.org) **The ALS Association (US)**
- [www.lesturnerals.org](http://www.lesturnerals.org) **The Les Turner ALS Foundation** (Chicago-based ALS organization)
- [www.march-of-faces.org](http://www.march-of-faces.org) **March of Faces** – awareness raising project
- [www.als-mda.org](http://www.als-mda.org) **Muscular Dystrophy Association (MDA) (US)** also supports people with ALS and funds ALS research
- [www.mnda.org](http://www.mnda.org) **Motor Neurone Disease Association (UK)**

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# RECOMMENDED READING AND OTHER RESOURCES

## ALS Research:

- **ALS Society of Canada** [www.als.ca](http://www.als.ca)
  - ▶ **ALS Society of Canada Research Updates**  
[Click Here](#)
  - ▶ **ALS Society of Canada Research Synopsis**  
[Click Here](#)
- **Canadian ALS Research Network (CALS)**  
[www.alsnetwork.ca](http://www.alsnetwork.ca)
- **The ALS Association (USA)** [www.alsa.org](http://www.alsa.org)
- **The ALS C.A.R.E. Program** is a voluntary, physician-directed program to improve clinical outcomes for patients diagnosed with ALS [www.outcomes-umassmed.org/als](http://www.outcomes-umassmed.org/als)
- **The Eleanor and Lou Gehrig MDA/ALS Research Centre** <http://www.columbiaals.org/>
- **The Robert Packard Centre for ALS Research at Johns Hopkins**  
<http://www.hopkinsmedicine.org/alscenter/index.cfm>
- **The World Federation of Neurology ALS Site**  
[www.wfnals.org](http://www.wfnals.org)
- **Clinical Studies** <http://clinicaltrials.gov> (conduct search by disease)

## ALS Drug: Rilutek™:

- **Manufacturer's site** [www.alsinfo.com/index.jsp](http://www.alsinfo.com/index.jsp).

## Palliative Care:

- **Canadian Hospice and Palliative Care Association** [www.chpca.net](http://www.chpca.net)
- **Council on Palliative Care**  
[www.med.mcgill.ca/orgs/palcare/copchome.htm](http://www.med.mcgill.ca/orgs/palcare/copchome.htm)
- **End-of-Life Physician Education Resource Center** [www.eperc.mcw.edu](http://www.eperc.mcw.edu)
- **Promoting Excellence in End-of-Life Care**  
[www.promotingexcellence.org](http://www.promotingexcellence.org)

# ALS SOCIETIES AND CLINICS

## **ALS Society of Canada**

393 University Avenue, Suite 1701

Toronto, ON M5G 1E6

Web site: [www.als.ca](http://www.als.ca)

Provincial ALS Societies ([Click here for contact information listed by Province](#))

ALS Clinics and Centres in Canada ([Click here for contact information listed by Province](#))