ALS SLA
CANADA

Margot Algie, living with ALS since 2015

CANADIAN BEST PRACTICE RECOMMENDATIONS
FOR THE MANAGEMENT OF AMYOTROPHIC LATERAL SCLEROSIS

ESTABLISHING A NATIONAL STANDARD FOR ALS CARE AND TREATMENT IN CANADA

AN OVERVIEW

February 2022

FOREWORD

In November 2020, the first-ever Best Practice Recommendations for the management of ALS were published in the Canadian Medical Association Journal (CMAJ). Developed over a number of years by a Working Group of Canadian ALS clinicians, the comprehensive document represents what these experts in ALS care agree should be the standard of care for any person in Canada diagnosed with the disease – no matter where they live in our country.

An important step forward, the recommendations were developed to help enable ALS clinics across the country to meet a common national standard, offering the best possible care to patients and their families and helping them navigate this complex and progressive disease. We hope this summary will help everyone affected by ALS understand the agreed-upon standards of care.

On behalf of the Working Group, thank you to Canadians coast-to-coast whose generous donations to ALS Canada through the Ice Bucket Challenge helped fund this initiative. We also extend our appreciation to the Canadian ALS Research Network (CALS) for its support of this effort, Federation partners for their critical feedback, those who provided coordination and logistical support, and most importantly, the people who shared their lived experience with ALS as we developed this important new tool.

Christen Shoesmith, MD, FRCPC
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Dr. Christen Shoesmith

"An important step forward, the recommendations were developed to help enable ALS clinics across the country to meet a common national standard, offering the best possible care to patients and their families and helping them navigate this complex and progressive disease."

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INTRODUCTION

Amyotrophic Lateral Sclerosis (also known as ALS, Lou Gehrig's disease or motor neuron disease) is a disease of the brain and spinal cord that causes gradual muscle weakness and eventually results in paralysis. People with ALS lose the ability to move and speak.

With four out of five people dying within five years of diagnosis in Canada, providing timely, optimal care from coast-to-coast is critical.

Until the recent publication of the Best Practice Recommendations (BPRs), there were no published guidelines explicitly for the care of people living with ALS in Canada. Health care providers had to rely on those published in the US and Europe, which do not necessarily reflect our unique experience, values and the health care system. This has meant that people living with ALS may have different experiences accessing care and resources, as well as local health system supports.

To address these gaps and provide an update on best practices, a 13-member pan-Canadian Working Group of experts reviewed evidence and enlisted the feedback and input of stakeholders across the country as they developed more than 130 recommendations across 13 areas of focus.

These BPRs are a comprehensive resource that:

Establishes
a national standard for the care and management of people living with ALS and addresses issues relevant to a Canadian context.

Enables
healthcare providers to offer the best possible care and help families affected by ALS to navigate this complex and devastating disease.

Empowers
people living with ALS, their families and healthcare providers to make informed, collaborative decisions about ALS – no matter where they reside in Canada.

Creates
a foundation to advocate for the best possible standards of care consistent with the best available evidence and expert consensus.

This summary booklet provides an overview of the BPRs, which underscore the importance of multidisciplinary care across all areas, which range from communicating the diagnosis of ALS, to symptom management to the importance of exercise. The full manuscript is available through CMAJ.

The BPRs are not designed to replace any clinician's or care team's management, as every person and family in their care is unique. Rather, they are designed to support decision-making and inform collaborative discussions between a person with ALS and their care team members. We hope this summary will help people diagnosed with ALS and their families understand the agreed-upon recommendations and empower them to advocate for the best possible standard of care.

DEVELOPMENT PROCESS

To develop the **Best Practice Recommendations,** the Working Group:

- Assessed hundreds of published peer-reviewed articles.
- Sought the input of people living with ALS in Canada, ALS Societies across the country.
- Engaged other health providers and organizations to share their opinion.

Where high quality published evidence was unavailable, the Working Group reached consensus based on their expertise and experience. For further detail on any aspect of the BPRs, including the research approach, tables, all references and citations, as well as notes as to the level of evidence and/or expert consensus for each recommendation, we encourage you to consult the complete article.

Adam Welburn Ross, living with ALS since 2018, with mother Gail Ingersoll and stepfather Gord Ingersoll

BEST PRACTICE RECOMMENDATIONS

In the published BPRs, the Working Group highlights that the care and management of patients with ALS should always be patient-focused, with attention to whole person and their emotional aspects of well-being.

The sections below highlight some of the key recommendations.

COMMUNICATION OF DIAGNOSIS

Patient needs and experiences during the diagnosis of ALS can be very different. Some may be overwhelmed with the news, yet others may feel they did not receive enough information at this critical time. The recommendations include the important concept of doctors tailoring the delivery of the diagnosis to the individual needs of their patient.

To do so, clinicians should seek to understand the feelings of their patient, address sources of information and community support, and provide timely follow-up after the diagnosis is first discussed. The Working Group also agreed:

- The diagnosis of ALS should be confirmed by a neurologist or physiatrist with training and expertise in ALS. Referrals for confirmation of an initial diagnosis of ALS should be seen in an ALS specialty clinic within 4 weeks.
- Discussion about ALS treatments and ALS research should occur. People with ALS should be provided with written information about ALS resources (paper or internet-based) and encouraged to register with their local ALS Society.
- Prognosis does not need to be discussed at the time of the initial diagnosis. However, these discussions are important and can be tailored to the needs of the individual.

MEDICATION ALIGNMENT

People diagnosed with ALS may already be on medications when they are first referred to a multidisciplinary clinic. Some may be for ALS symptoms, and others for unrelated health issues. The clinic team will review all medications at first visit and will regularly assess treatments to ensure all are necessary.

MULTIDISCIPLINARY CARE

People living with ALS who are followed by a multidisciplinary clinic have been shown to have better outcomes, including improved survival, fewer hospital admissions, increased use of adaptive equipment and enhanced quality of life.

That's why people with ALS should be referred to specialized ALS clinics. This care should be delivered through a team-based approach, with physicians and other health professionals addressing multiple care issues. This includes communication, nutrition, swallowing, mobility, activities of daily living, respiratory care, cognition, psychosocial issues, medical management and end-of-life care. Care should also be a collaboration between the family physician and the ALS clinic, with staff available for remote consultation between patient visits.

The frequency of clinic visits is dictated by the needs and rate of progression of the person with ALS. A dedicated nurse or other clinic allied health care professional should be available to support people with ALS and their family members to discuss issues between clinic visits.

This underscores the growing and vital role of telemedicine and telehealth resources. Virtual care can supplement clinic-based care by proactively monitoring the health of the person living with ALS – potentially helping prevent complications.

CAREGIVERS

ALS impacts both the person diagnosed with the disease, as well as informal caregivers, such as family members or friends, who play a valuable role.

Many people with ALS worry about possible burdens on their loved ones. Health care providers, therefore, need to consider the physical and emotional well-being of caregivers, and involve them in planning for the impact of ALS on both the person with ALS and themselves, and provide information on existing relief programs where possible.

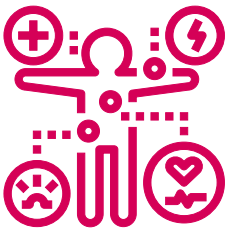
Assessing caregiver burden, coping strategies, mood and family dynamics would help identify caregivers and families in need of respite and support services. Local ALS societies may also have resources for family members and caregivers.

DISEASE-MODIFYING THERAPIES

At the time of publication in November 2020, there are two Health Canada-approved treatments for ALS in Canada – riluzole and edaravone. Both are known as '*disease-modifying therapies*', which work to try and slow the disease process. They are different from medications used to treat a symptom of the disease, such as muscle cramps.

Although the therapies currently available are not cures, research studies have shown that they can help in many people with ALS. Disease-modifying treatments should only be prescribed by clinicians with experience managing patients with ALS. People with ALS and their health care provider are encouraged to have an open dialogue about their potential risks and benefits of therapies, including those not approved for the treatment of ALS.

SYMPTOM MANAGEMENT



People living with ALS often have multiple uncomfortable or bothersome symptoms that interfere with function and quality of life. This can include pain, muscle twitches (fasciculations), drooling (sialorrhea), impaired emotional control (pseudobulbar affect), muscle stiffness, (spasticity), cramps, depression, anxiety, insomnia and fatigue.

The wide range of symptoms underscores the importance of access to and ongoing consultation with a multidisciplinary care clinic for appropriate monitoring and management. Most of these symptoms have treatments to reduce their severity, sometimes through medication or other types of non-drug therapy. The person living with ALS and their doctor should discuss the risks and benefits of all medications and therapies.

Some of the specific recommendations to manage symptoms, based on evidence where available as well as expert consensus, include:

PAIN

- Pain is a recognized aspect of ALS, with many potential causes. Symptoms should be regularly assessed, and treatments tailored towards the specific cause.

MUSCLE TWITCHES (*fasciculations*)

- In most people living with ALS, fasciculations do not need to be managed with medication.
- If they cause substantial distress, the medication gabapentin can be considered.

Peter Wood, living with ALS since 2015, and daughter Siena

EXCESS SALIVA (*sialorrhea*)

- Multiple therapies are available to address excess saliva, including medications in the anticholinergic family and botulinum toxin.
- Oral suction can be used as a management option.

IMPAIRED EMOTIONAL CONTROL/EMOTIONAL LABILITY (*pseudobulbar affect*)

- Impaired emotional control or emotional lability (difficulty controlling laughing or crying) is a symptom of ALS and is not always associated with depression. Dextromethorphan combined with quinidine is one of the treatments that can be used to treat emotional lability. However, treatment for emotional lability is not required unless it's distressing to the person with ALS.
- Medications are also available to treat concurrent symptoms in addition to the emotional lability, including amitriptyline for sleep, or a selective serotonin reuptake inhibitor (SSRI) for depression.

TIGHT LIMBS (*spasticity*)

- Range of motion exercises, often prescribed by a physiotherapist or occupational therapist, can be useful for managing spasticity.
- If medication is required, a number of options may be considered, including baclofen, tizanidine, botulinum toxin, and benzodiazepines. Cannabinoids could also be considered.

CRAMPS

- Muscle cramps need to be differentiated from other causes of pain.
- First-line management could include tonic water, or the medications gabapentin and baclofen.

DEPRESSION & ANXIETY

- Depression and anxiety should be treated in ALS, since both have a substantial impact on well-being.
- Depression and anti-anxiety medications may be prescribed, in addition to other supports, such as psychology, social work, psychiatry or spiritual care.

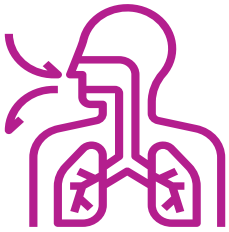
INSOMNIA

- There are many causes of sleeplessness, such as respiratory difficulties and depression that should be appropriately investigated.
- Management of insomnia will depend on the cause.

FATIGUE

- Reversible causes of fatigue should be considered, such as respiratory insufficiency, sleep disorders, depression, medication side effects, and riluzole use.
- An occupational therapist may discuss techniques to conserve energy.

RESPIRATORY MANAGEMENT



Breathing muscles help with the important functions of breathing and keeping airways clear. One of the important aspects of having access to a multidisciplinary care clinic is regular monitoring of respiratory signs and symptoms, in addition to ongoing adjustments and troubleshooting of equipment and devices that support breathing.

Non-invasive ventilation (NIV)

Non-invasive ventilation (NIV) involves using a mask attached to machine to help one breathe. A BIPAP device is a common form of NIV. NIV is the standard of care to treat respiratory insufficiency in ALS, both to lengthen survival and manage symptoms, as well as enhance quality of life.

The Working Group agreed people with ALS should start NIV when they have symptoms of respiratory impairment. Those without respiratory symptoms should start to use NIV when their Forced Vital Capacity (the amount of air expelled from the lungs as measured by a test) is at 65%. There are also other criteria for starting NIV detailed in the BPRs. If criteria are met, NIV should be started within 4 weeks.

Oxygen Use

Oxygen should not be considered a routine treatment for breathing difficulties. In patients with ALS with low blood oxygen levels, NIV needs to be considered first. If low blood oxygen levels remain with the use of NIV, assessments should be made and supplemental oxygen can be considered.

Managing secretions

Secretions like phlegm and mucus can be distressing to people with ALS, especially when a person's cough strength is weak. The ALS clinic may recommend some medications, equipment and exercises that can help keep airways clear.

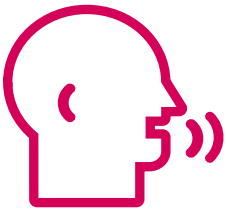
Therapists or nurses at the ALS clinic will help educate people with ALS so they are comfortable with devices and these experts will modify settings through ongoing monitoring and consultation.

Support for respiratory equipment

The Working Group agreed that providing adequate in-home respiratory support for NIV and machines to assist coughing is essential, regardless of whether the patient resides in their own home, long-term care facility or hospice. Support can include education, machine adjustments, and troubleshooting.

Dr. Jeff Sutherland, living with ALS since 2007

SPEAKING PROBLEMS (*dysarthria*)

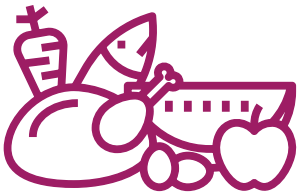


The ability to communicate thoughts and needs to others is vitally important to individuals. ALS often impairs the ability to talk. There are many tools to support communication, including low-tech options, such as letter- or picture-boards, as well as high-tech options, such as use of tablets, voice amplification and voice banking.

Providing access to different types of communication supports can allow independence, participation and better quality of life. This may also benefit caregivers.

As people with ALS experience loss of function, some modes of communication may no longer be possible. That's why people with dysarthria should be regularly followed by speech language pathologist to ensure timely interventions. Devices should be offered to eligible people with ALS in early disease stages. Those in later stages will also benefit from communication devices and strategies.

NUTRITIONAL MANAGEMENT



Ongoing nutrition management is very important in ALS. People living with the disease need extra calories because they burn them at a high rate. Generally, an appropriate member of the multidisciplinary team, such as a dietician, should follow weight and monitor nutrition every 3 months. They will also provide guidance, including adjustments to diet, such as high calorie approaches.

People with ALS should also be followed by a certified swallowing clinician who will monitor swallowing safety. They may recommend changes to the texture and consistency of food and liquids that are consumed. The multidisciplinary team may suggest a feeding tube for nutrition and/or hydration.

A feeding tube is soft and flexible, and is inserted through the belly into the stomach (often in a day procedure by a surgeon or radiologist). It supplements and supports nutrition, hydration and helps deliver medication. It's important to know that people with ALS who have a feeding tube will often also eat by mouth. There should be regular support by a registered dietician to follow and advise on the nutrition delivered through the feeding tube.

EXERCISE

Research suggests that exercise in ALS is beneficial for function and quality of life. There is no demonstrated harm from exercise. A personalized exercise program, including strength and aerobic training, should be encouraged for people with ALS who are able to participate. A regular stretching and range-of-motion program is recommended to manage spasticity and pain, and prevent shortening and stiffening of muscles, tendons, or other tissue.

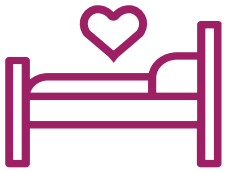
COGNITION AND BEHAVIOUR

In some people with ALS, there may be a change in behaviour, personality or ability to problem solve. At this time, there are no effective drug treatments for cognitive or behavioural impairment in ALS. However, a multidisciplinary approach can be considered to manage behaviour issues.

Screening for cognitive and behavioural impairment should be performed in people diagnosed with ALS early in their disease. However, if there is concern about cognition or behaviour at any point, specific assessments can occur with the person and their family members or caregiver.

A multidisciplinary approach can be considered to manage particularly problematic behaviours. Involving a behavioural specialist (such as an occupational therapist or psychologist) or psychiatrist for assistance may be considered.

PALLIATIVE CARE



Palliative care is care given to improve the quality of life of patients who have a serious or life-threatening disease, such as ALS. Palliative care is an approach that addresses the person as a whole, not just their disease.

Advanced care planning is also an important aspect of ALS management because it helps establish a person's preferences before the disease is advanced and communication is impaired. These conversations should be started early in the disease or whenever the person with ALS inquires. Ongoing discussions about goals of care should be part of routine follow-up, and throughout the disease course. They can be had with ALS clinic staff, palliative care practitioners and family physicians.

Health care providers are advised that conversations about goals of care should be introduced according to the person's readiness and style of decision-making. Considerations should include preferred breathing and nutrition supports, tracheostomy (an airway created in the neck), and other life prolonging or comfort care measures.

Some people living with ALS may wish to discuss medical assistance in dying (MAiD) and should do so with their physician or nurse practitioner, who will be knowledgeable of regional guidelines. Regardless of decisions related to MAiD, people living with ALS must have access to the appropriate palliative care to meet their needs.

The person with ALS should also discuss their preferences about end-of-life care with family members and caregivers. This may include the subject of organ donation, as people with ALS may be accepted as solid organ donors (as determined by their local organ donation organization). People with ALS interested in learning more about organ donation should ask their health care team.

While there are many facets to palliative care, it's important that people living with ALS in Canada can access the same basic supports - no matter where they reside.

ALS SLA CANADA

Our goal in developing these recommendations was to help enable the dedicated staff and team members of ALS clinics across Canada to meet a common national standard, offering the best possible care to families and helping them navigate this complex and progressive disease.

We also believe this important new tool will empower people living with ALS, their families and their health care providers to advocate for access to optimal care and treatment according to these best practice recommendations – no matter where they live in Canada.

BEST PRACTICE RECOMMENDATIONS FOR THE MANAGEMENT OF ALS WORKING GROUP

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