

XXXX

British Columbia

Living with ALS

1. Are there any recommendations you do not agree with? Please be specific about what change(s) you would like to suggest to improve the statements.

Line number (or span)	Comments
Disease-modifying Therapies 6c	Notwithstanding the cost of the drug. Given that its benefits are somewhat minimal, I think it is a personal decision as to how one should spend their money at the time of diagnosis. If a drug is completely covered by insurance and recommended by doctors then it should be pursued, sure. Otherwise, there may be more proactive uses for one's money at this time and going forward.

Noted

2. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
Communication of diagnosis #3	Given the nature of the disease, it is easy for patients to misunderstand treatments from third parties that are marketed as potentially beneficial, but are essentially scams. Perhaps the written information provided to a ALS patient can provide an explanation that only peer-reviewed and approved treatments should be researched for consideration, or at least caution the patient that the scams do exist. Noted, no changes as will be discussed by ALS MD
Disease-modifying therapies #8	I think a very pragmatic approach should be had when recommending disease-modifying therapies for patients. For instance, pursuing Edaravone for potentially limited benefit, at the start of the disease progression cycle may not be the most intelligent choice for all patients. Instead of being tied to regular infusions and exorbitant costs at the start of your disease, perhaps there is a better usage for one's time and resources. This is a personal choice that I think every patient needs to consider. At the early onset of my disease, I spent a lot of my time and money traveling. I do not regret this decision, even in hindsight, knowing that treatments like Edaravone were possible. NOTED and will be discussed individually

3. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

General Comments
Obviously this document was written for medical professionals. The vocabulary and jargon in the article is not easily understood by the average patient, but I believe that is as per the design of the document. Lay person communication planned with roll out.

4. Is there anything that you would suggest to make these recommendations more useful or user friendly for you or your clients?

Line number (or span)	Comments
Communication of diagnosis #4	Although the disease is undoubtedly terminal, I found that during my diagnosis it was particularly comforting to know that changes in my physical well-being we're not going to be instantaneous. As traumatic as receiving the diagnosis is, knowing that I wasn't going to drop dead tomorrow, for instance, gave me pause to think before simply reacting to the terrible news I was receiving. "You are going to die" is a very different message to receive than "you're going to die, eventually", even though both are true. Addressed by delivering info in chunks and individually tailored.

XXXXXX

British Columbia

Caregiver

5. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
CAREGIVERS	ASSISTANCE IN TRAINING HOW BEST TO ASSIST WITH; MOVEMENT, TRANSFER BED-CHAIR, CHAIR-TOILET, ETC. Addressed via multidisc care
PALLIATIVE CARE 108 - 110	ARE THERE OTHER 'CARE OPTIONS' AVAILABLE IF THE ALS PATIENTS NEEDS BECOME GREATER THAN CAN BE PROVIDED FOR AT HOME? Will be regionally dependent, and introduced via community palliative care

6. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

Line number (or span)	Comments
131	ABBREVIATIONS

General Comments
THERE ARE ABBREVIATIONS THAT DO HELP THE READER, HOWEVER THEY WERE FOUND ON PAGE 8/11 PAGES. I CUT AND PASTE TO A SEPARATE DOCUMENT BEFORE RE-READING, IN AN ATTEMPT TO FULLY UNDERSTAND THE CONTENT. Leave to CMAJ to decide

7. Is there anything that you would suggest to make these recommendations more useful or user friendly for you or your clients?

General Comments
UNDERSTANDABLY, THIS DOCUMENT CONTAINS REFERENCE TO MEDICAL CONDITION AND/OR TREATMENT THAT IS NOT CLEAR TO THE LAY PERSON.

ALS Clinic Team allied health: OT, RD, SLP, RN, SW

British Columbia

Healthcare professionals

8. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
Respiratory Management – Screening line number 15	Talks about having respiratory baseline, but does not indicate when the baseline respiratory status should be taken. Often the timing on when this is done is asked by clients. Perhaps a statement clarifying this would be useful. Baseline is first clinic visit

Recommendations – Medical Assistance in Dying – line number 123	Add Social worker MD or NP needs to be ultimate person discussing.. Won't add SW since this may be provincially variable

9. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

Line number (or span)	Comments
Nutritional Management – Supplements/Enteral Feeds line 45-46	Currently you're talking about high calorie diets and parenteral nutrition. However the title implies that the recommendations are around oral nutritional supplements and enteral nutrition. Consider changing the title from "Supplements/Enteral Feeds" to "Diet Modifications and Nutrition Support" or something similar. No comments made around home blenderized tube feeds. Changed
Palliative care – treatments - Line number 111	Suggest to delete " for a good death" to reduce redundancy and confusion. Ending with "...versus comfort focused care" already implies the latter. We feel its important to keep that phrase in as a prompt to discuss death and dying.
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Saskatchewan

Client Services Consultant for ALS Society

1. Are there any recommendations you do not agree with? Please be specific about what change(s) you would like to suggest to improve the statements.

Line number (or span)	Comments
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#76-77	On page 6 in the Depression section, it should make mention of referring clients to non-pharmacological supports as it does in the anxiety section. Psychosocial support is important for someone struggling through depression especially through a terminal illness. Added
#83-85	On page 6 in the Insomnia section psychosocial supports for client should be discussed as well. It does state that Insomnia can be a symptom of depression, but only mentions pharmaceutical management if depression is a part of the cause of insomnia other psychosocial support should be mentioned. Will now be covered via referring back to depression
#86-87	On page 6 in the fatigue section it discusses adjusting medication to offset fatigue, but it does not discuss how to manage if the cause of fatigue is not medication related. I think there should be a section stating, "consider non-pharmaceutical options if the fatigue is depression related". No evidence to support this

2. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
#105-107	The discussion of anticipatory grief, and grief and loss that caregivers are likely going through. That they be monitored for this and are aware of what this may look like. Have them connected to psychosocial supports.
	The discussion of grief and loss for clients. They are likely grieving throughout the disease process in various ways and need to be aware of this. Should be connected to outside resources that could benefit their psychosocial well-being. We are assuming that grief will be included within emotional well-being needs.

3. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

4.

General Comments
I think it is straight-forward and easy to understand.

5. Is there anything that you would suggest to make these recommendations more useful or user friendly for you or your clients?

General Comments

If our clients will be utilizing these – perhaps a definition section, as there is an abbreviation section. **Lay person summaries will be separate**

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Quebec

ALS Society Employee and Healthcare professional

1. Are there any recommendations you do not agree with? Please be specific about what change(s) you would like to suggest to improve the statements.

General Comments

Caregivers: There should be an expansion of content and practice recommendations.

The nature of caregiving in general and ALS in particular is complex; caregivers are the experts of what’s going on in the day-to-day and in general want to be included in care planning and have their expertise recognized and valued. At the same time, their primary role is that of spouse, child, other family member, friend, etc.... Caregivers should be given the opportunity to determine if, when, how, and in what way they will contribute to the care of the person with ALS, free from undue pressure. This type of check-in should be done regularly for the caregiver’s wellbeing, especially when changes in the care plan might require their involvement. This way, should they express discomfort at the idea of being involved, alternative solutions can be found.

Ideally, a patient file would also be opened for at least the primary caregiver to allow for social services, spiritual care, psychology, and others to provide services.

We tried to address this globally since we are stating that we are trying to involve them in the process

2. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
Recommendation 2	“Timely clinical contact”; would suggest defining “timely”. This had been discussed at length, and we couldn’t agree on an interval and requires individualized patient attention
Recommendations 6 & 7	No mention of conversations around possible side effects and

Recommendation 21	likelihood of occurrence of side effects. This is an automatic discussion re prescribing any medication via the MD
Recommendation 30 but also most major interventions	In what ways might NIV change the trajectory of the EOL experience? Will consider
Palliative Care	Patients need to be made aware that it is their right to withdraw care at any time, i.e., remove the vent, stop feeds, etc.... In my experience, patients often have the impression that once they opt for a trach with invasive ventilation, for example, they are resigned to that fate. Without trivializing the decision to withdraw care, patients need to have all of the relevant information at their disposal to facilitate informed decision-making. We have added a sentence to address this.
Missing	Explanation of palliative care as care without curative intent/symptom management so that early involvement of palliative care does not cause undue anxiety to families. The full palliative care supplement document has the definition. Driving assessments/adaptations Not included in our literature review

General Comments
Multidisciplinary care: In our province, we have 3 confirmed ALS clinics (as defined by offering multidisciplinary care and research), with a 4 th coming online. Many patients live prohibitively far away from these clinics. The committee mentions telemedicine and telehealth as options for supplementing clinic-based multidisciplinary care but it would be good to also mention working with social services to find ways to improve or facilitate access to multidisciplinary clinic care, e.g., transportation, financial aid, etc.... Hard to put in guidelines

3. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

General Comments
It is difficult for me to comment on this without seeing the version intended for the lay-person. As a social worker with hospital experience prior to my role at the ALS Society, I understood much of the content but the very detailed information on breathing or nutrition metrics were not accessible to me.

4. Is there anything that you would suggest to make these recommendations more useful or user friendly for you or your clients?

General Comments
I'm looking forward to seeing the versions intended for laypeople.

XXXXXX

Quebec

Board member, caregiver, nurse and project manager (also listed healthcare professional)

1. Are there any recommendations you do not agree with? Please be specific about what change(s) you would like to suggest to improve the statements.
- 2.

General Comments
<p>Question 1:</p> <p>Caregivers: There should be an expansion of content and practice recommendations. What was stated was based evidence review, space is limited. Caregiver inclusion is new for any guidelines</p> <p>Beyond being 'involved' in planning for the impact of ALS. There is a need for Healthcare providers to engage caregivers actively in discussions pertaining to their role, ability and willingness to be involved in changes in level of care and the support required for the patient with ALS. Caregivers are partners in the care plan for the patient with ALS. As such, discussions pertaining to changes in the treatment plan should include evaluation of the impact on the health and wellbeing of the caregiver. Assessment of the requirement for additional supportive resources that may be necessary to enhance the health of caregivers/family members should be carried out when any significant treatment change is anticipated or necessary. Already included in the general statements</p>

3. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
Medication alignment #52-56	There is no mention of evidence relating to the place or use of cannabinoids for pain management in ALS (there is one mention under spasticity (71) but no mention under pain(57-58). There is no evidence for cannabis in ALS (yet)

Symptom Management	There is no mention of xerostomia symptoms in ALS patients, nor recommendations or evidence related to symptomatic management. This has not been evaluated in the literature yet
Nutritional Management	Minimal information related to the role of the dietician-nutritionist within the ALS team in the education and supportive care of patients with ALS who have decided to pursue enteral feeding. (No mention of impact on caregiving) No recommendations or evidence related to enteral formulas versus blenderized diets. Dietitian: This is implied by the high level nutritional section
Organ Donation	There is no content regarding patients with ALS who have chosen MAID and wish to be organ donors. Not necessary to clarify
Figure 2: Nutritional Management	Under the intervention column(category) there is no option or alternative to consider palliative care options. This gives the impression that all patients with ALS undergo RIG, PEG, NG or Parenteral nutrition. Every intervention is optional, we don't have to be explicit about this for every option. "Consider" means its option.

General Comments
<p>Question 2:</p> <p>There is no recommendation or information regarding the potential importance of oral hygiene and ensuring ongoing collaborative dental care for ALS patients. There are a number of studies which address oral hygiene and its impact on aspiration pneumonia in ALS patients that do and/or do not have a feeding tube inserted, and those who have both non-invasive and invasive ventilation assistance. This question was not researched during the guideline development. It is a great suggestion to consider this for future guideline revisions.</p> <p>Table 3: does not include Xerostomia (not researched during guideline development)</p>

4. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

Line number (or span)	Comments
Respiratory management recommendations #17	Format error: The sub-bullet point 'i' should be deleted only the * should remain. Done
Recommendations #20a & 34a	Is this statement true for all provinces across Canada? Yes

Supplements/Enteral feeds # 45	There is no content, information or evidence recommendation relating to high-calorie/high-protein diets (link to prevention of muscle wasting and increased caloric requirements in ALS) This is included in the supplement nutrition discuss, but the evidence wasn't as high as the high cal/high carb diet.
Pain # 57 -58	It may be worth mentioning that causes of pain are multifactorial and can stem from physical, psychological, spiritual and existential spheres and that all of these are to be assessed on an ongoing basis across the course of the illness. This can be implied by the "many potential causes". Since we have to be concise, we cannot expand on all points. The potential causes of pain are widely reference in pain literature.
Recommendation Pseudobulbar effect #68	Format error: opening bracket is missing ; EC) Done.
Anxiety # 80	Rephrasing suggestion: If depression is concurrently present, an SSRI should be prescribed (EC). Changed
Fatigue #86	Consider the addition of reversible causes: malnutrition, dehydration, infection. We cannot list all potential causes and have given a few examples which are common in ALS patients Referral to allied professionals such as occupational therapist for possible energy conservation measures with ALS'S (? level of evidence) Reasonable suggestion
Dysarthria #93	Rephrasing suggestion: Voice banking may be offered to eligible patients in early stages of speech impairment (EC). We are comfortable leaving as is.

General Comments
For Question 3: Under Organ donation: #127: consider removal of the word solid (ie: solid organ donors versus organ donors). Solid organ terminology used by organ donation organizations (CS to check)

5. Is there anything that you would suggest to make these recommendations more useful or user friendly for you or your clients?

Line number (or span)	Comments
Feeding tube insertion # 43	Consider rephrasing: Once a tube feeding is placed, an experienced clinician (endoscopist or radiologist) is readily available to address immediate and latent tube complications (EC). We prefer original wording

Feeding tube insertion	I would recommend as possible content which defines the standard of care and follow up timeframe once a feeding tube is inserted. (Role of nutritionist/ALS team, nutritional information, choice of enteral formula, interval of assessment and troubleshooting) According to the current information it would be every 3 months or as clinically indicated. Good suggestion. We have added this to point 43.
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General Comments
<p>Question 4:</p> <p>I would recommend a legend for Table 2 specific to the Grade recommendations or Evidence Levels so that the reader is clear regarding the significance of EC, Level A, Level, B and Level C. This would make the recommendations clear for all categories of reader. This will be in the body of the manuscript.</p>

Former caregiver from Quebec

Translation : I read the attached document and even had the chance to discuss with the nurse clinician, Sophie Villemaire, at the CHUM last Thursday. I don't have any comments to add, for me the document is clear and covered all subjects.

XXXXX, SLP Alberta

Line number (or span)	Comments – <i>Potential changes for consideration</i>
3	<ul style="list-style-type: none"> - Changed to ensure accessibility of information <p>Discussion about ALS treatments and ALS research should occur. Patients should be provided with written information about ALS resources (accessible to the individual's level of cognitive-communication capacity) and community support organizations (EC). Including this would be too wordy. We do rely on the clinicians to provided appropriate cognitive level/accessible reading material</p>
5	<ul style="list-style-type: none"> - Changed to patient-first language <p>Disease-modifying treatments should be prescribed by clinicians with experience managing ALS patients patients with ALS (EC). Changed</p>

11	<p>- Nursing is distinct from allied health</p> <p>Multidisciplinary care should be delivered through a team-based approach, with physicians, nurses and allied health professionals addressing issues including communication, nutrition, swallowing, mobility, activities of daily living, respiratory care, cognition, psychosocial issues, medical management, and end-of-life care (EC) We will change to “other health professionals”</p>
13	<p>- Nursing is distinct from allied health</p> <p>A dedicated nurse and other clinic allied health care professionals should be available to support patients and their family members for ALS issues between clinic visits (EC). We were mostly adding this point for a “point person” to coordinate care between visits. They can help navigate issues between visits (even referring to allied health professionals)</p>
15	<p>- Patient-first language</p> <p>ALS patients Patients with ALS need regular respiratory monitoring at baseline and every 3 months, or as clinically indicated (EC). Regular respiratory monitoring should include:</p>
37	<p>- Compensatory recommendations (positional, strategies, etc.) are separate from consistency modifications and are an important part of care.</p> <p>Nutritional interventions, including dietary alteration and consideration of referral for enteral tube insertion, are indicated at diagnosis or at follow-up if there is: (1) increased risk of aspiration despite consistency modifications and compensatory recommendations (EC)(2)≥5-10% reduction in weight from usual or baseline weight (level C); (3) ≥1-point reduction in BMI from usual or baseline BMI (level B); (4) BMI <18.5 (level B); or (5) TDEE exceeds daily energy intake (EC). Added</p>
49	<p>- Patient-first language</p> <p>There is no evidence to suggest screening for thromboembolism in asymptomatic ALS patients patients with ALS (EC).</p>
51	<p>- Patient-first language</p> <p>If VTE occurs in an ALS patient a patient with ALS, they should be anticoagulated as per standard VTE guidelines (EC)</p>
88	<p>- Inclusion of assessment</p> <p>Patients with dysarthria should be regularly followed by SLP to ensure timely communication assessment and interventions (EC). Since we imply that interventions require assessment, we are OK leaving as is</p>
89	<p>Use of augmentative and alternative communication devices should be offered to eligible patients in early disease stages (EC). Patients in later disease stages will also benefit from communication devices and strategies (EC). Agree</p>

93	<p>- Wording change recommendation</p> <p>Voice banking may should be offered to appropriate patients (EC). Agree</p>
105	<p>- Integration of palliative care guideline components</p> <p>Health care providers should be attentive to the needs, preferences and emotional well-being of caregivers, and assess caregivers’ skills and need for training and support, to assist with care of a patient with ALS. Caregivers should be involved in planning for the impact of ALS on both the patient and themselves (EC). We are starting with basic statements about caregivers first, as this is the first inclusions in guidelines.</p>
107	<p>- Consideration of palliative care guideline: “Provide appropriate guidance, support and information to families and caregivers based on awareness of culture and needs, and directs them to bereavement services as required”</p> <p>Assessment of caregiver burden, coping strategies, mood, family dynamics, cultural and spiritual beliefs, and needs would assist in identifying caregivers and families in need of respite, supportive and bereavement services as required (EC). Word count is too high to include this in both locations</p>
108	<p>- Palliative care guidelines indicate: “A palliative approach to care starts early in the trajectory of a progressive life-limiting condition, and may be appropriate at the time of diagnosis”</p> <p>- Inclusion of other professions in a palliative care approach</p> <p>Palliative care of patients with ALS can be provided throughout the disease course can start early in the trajectory of ALS progression, and may be appropriate at the time of diagnosis. Palliative care can be provided by ALS clinic staff, nursing, allied health teams, palliative care practitioners, and family physicians (EC). Need to be succinct and limit words.</p>
116	<p>- Incorporating palliative care guidelines.</p> <p>Patients should be encouraged to discuss their preferences about end-of-life care with family members and caregivers (EC). Health care providers should demonstrate openness, sensitivity and acceptance of social, spiritual and cultural values and practices that may influence patient and family preferences regarding end-of-life care. Need to be succinct and limit words.</p>

General Comments

Patient-first language was inconsistently used throughout the document (i.e., calling someone a “patient with ALS” or “person living with ALS” rather than an “ALS patient”). Patient-first language should be used consistently in healthcare guidelines.

1. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

General Comments

In reviewing the guidelines and discussing components with my supervisor, I thought it would be beneficial to mention of the role of genetic counselling and testing given the rates of familial ALS. My supervisor, Dr. Gerald Pfeffer, would be willing to provide a section on genetics in ALS, if the committee believes this would be helpful to include. **We agree. We had discussed this in advance and had decided not to include genetics at this time. We will include in subsequent revisions.**

It may also be beneficial to include guidelines regarding indigenous populations and equal access to care for people living in rural and isolated Northern regions of Canada. **We agree. The guidelines can be used to advocate for care for all patients.**

2. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

General Comments

Overall, the guidelines were very thorough and well written for understanding.

XXXXXX, Executive directive SK ALS

1. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
Recommendation 107/116	Should also include a path towards end of life preparation for the caregivers. The one thing that most of our caregivers say they wish they had done was prepare more for the end and know what resources they had access to in preparation for that time. Agree. This preparation will be included in palliative care and preparation for dying is a component of routine palliative care (we don't wish to restate in caregiver section).
Recommendation 3	Would it not be appropriate to indicate the local ALS Society where applicable. Great point. We have altered this sentence.
Recommendation 105/106/107	Again an indication of that the local ALS Society (where applicable) can play a role in helping to support the caregivers would be appropriate. Good suggestion

General Comments
<p>Great tool for reference for organizations like ours to better understand the medical side of things that we do not touch yet are influenced greatly by as we also accompany the patient/client through his or her ALS journey.</p> <p>No where in the tool do I see reference to the ALS Guide that each patient should receive when diagnosed. How and where to include that is unclear for me. This is implied by "ALS resources" in point 3.</p> <p>Lastly, I don't know if this is allowable ethically but at any one time I don't know how many ALS studies or research studies are underway in Canada do we want these to be shared with active clients so that they may contribute of themselves towards these endeavors. Maybe not appropriate or maybe not appropriate here but definitely worth discussing. This is part of the diagnostic discussion</p>

XXXXX, Ontario Patient

Line number (or span)	Comments
Communication of diagnosis	In the discussion in which the diagnosis of ALS is received by the patient, it feels akin to "drinking from a fire hose." It's both overwhelming and devastating. While it's important to convey that there are some treatments available for some situations, and that there are clinical trials and many research projects underway, I would urge caution and not contributing to the patient being overwhelmed. For many people receiving the diagnosis, it may be sufficient for instance to say that treatments and research will be discussed in a follow-up appointment. This is discussed

Communication of diagnosis	I and my fellow pALS (N = 3) believe that an immediate referral to both an ALS Clinic and the ALS Society are vitally important, and that agreement to do so ought to be obtained during the diagnosis discussion. The ALS Clinic is important for immediate follow-up. The ALS Society is important so that a rep can visit with the patient as soon as possible to help navigate through all of the information available and choices to be made, and to start understanding and navigating the myriad supports, which are available and yet which are not integrated well. Addressed in point 3
Communication of diagnosis	Where the communication of diagnosis is done by other than the family physician, it's important to encourage the patient to make an appointment and to go and talk with their physician as soon as possible. From talking to others, it would seem this doesn't always happen. The individual's physician will continue to quarterback the patient's care, especially with respect to the inevitable comorbidities that arise. We agree. We will address this in a subsequent revision.
Communication of diagnosis	it's important to recognize that the patient has a circle of loved ones with whom the diagnosis will be shared. Via referral, a quick connection with the ALS Clinic, ALS Society rep, or family physician is important in helping the patient deal with that very difficult sharing of the diagnosis. Connection to society added to point 3.
Disease-modifying therapies	I think it's important to recognize that as soon as this section is published, there is a high likelihood that it will be out of date. I'd suggest it's important to state explicitly in the guidelines that the accelerating pace of research and clinical trials heightens the need to stay current in this area. We also hope there will be significant changes in the future. Plans for revisions are in the manuscript body.
Disease-modifying therapies	With respect to #8 – this may be also the place to encourage ongoing dialogue with patients about research and clinical trials. Included in point 3.
Multidisciplinary care	I and my fellow pALS (N = 3) would like to underscore the vital importance, in our experience, of the ALS Clinics in providing an integrated multidisciplinary approach. As outlined in #11, the integrated and team-based approach is far more effective for we pALS, than an indeterminate sequence of separately booked visits with specialists without a quarterback to help pull it all together. We agree!
Symptom management	In reading through this section, it strikes me that the emotional and mental well-being of the patient is insufficiently served. It feels like #82 – non-pharmacological supports such as offered through psychology, social work, psychiatry, or spiritual care – are more important and perhaps deserve a subsection of their own. Attitude and emotional well-being provide a significant

	<p>component of quality of life, and perhaps need to be better recognized and supported. We have added extra supports.</p>
Caregivers	<p>Thank you for including this section. This is so important. The obvious aspects of ALS aside, the health and well-being of the patient and their caregiver(s) are tightly linked.</p>
Palliative care	<p>I would like to see this section expanded to address quality-of-life considerations rather than just end-of-life considerations. The availability and depth of palliative care resources varies from community to community within Ontario, and across the country. Given that palliative care is about quality of life, rather than just end-of-life care, I might suggest it's important for the patient and caregiver to understand the resources available in their area, how to contact them, and how to engage some of the services earlier than one might otherwise think needed. Discussion of advance care planning/directives, MAID, and other important aspects have a very large social/emotional component beyond the clinical aspects. As well, the ALS disease process is one in which the patient and caregiver are confronted with a steady succession of losses (of physical capability) and a concomitant steady succession of grieving cycles and an ever-increasing onslaught of anticipatory grief. Social workers, bereavement coordinators, and other therapists are important in helping to deal with this. It strikes me that recognizing this and giving it some visibility in the guidelines might be appropriate. Will plan to include this in the palliative care background document. (and may consider to have this in the main manuscript).</p>
Figure 1 & 2	<p>I suggest it might be appropriate to add to this figure some guidance on monitoring and intervention with respect to emotional/mental health. The figure as constructed would seem to be complete from a physical perspective, yet the emotional needs of the patient are excluded. These two figures were meant to be succinct with respect to respiratory care and nutritional management. We will include a statement in the manuscript that care providers should always practice holistic care including management of emotional and psychological well-being.</p>
Figure 3	<p>to reiterate some comments from above, non-pharmacological supports for emotional well-being are perhaps more important in more places than indicated, and</p>

	I might suggest giving that approach more visibility. Altered.
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XXXXXX, RT, BC

1. Are there any recommendations you feel that are missing or not addressed? If so, what are they?

Line number (or span)	Comments
18.	A respiratory specialist should also briefly discuss the possibility of invasive ventilation for a specific population. Just a soft introduction to the topic. It is not realistic to talk about NIV and not IV. If the respirologist is not going to bring this up at this juncture, it is extremely hard to field questions down the road in the community. I notice many don't bring this up. If you include this for recommendation #18, it may be helpful for people to plan and to understand that management of NIV is key in avoiding escalation of invasive ventilation. It provides some context. We do touch on IV in point 28 and 30. We don't necessarily discuss IV at the initiation of NIV. We discuss it at an appropriate time based on the patients needs and trajectory.
32.	Order of implementation of NIV and airway clearance. Some clarity on that. If airway clearance is a difficulty, NIV should be provided together. Often we provide a suction unit, and the ALS patient thinks that this is "enough for now", leading to urgency to NIV initiation down the road and stress for families. We suggest that both breathing and airway clearance functions should continue to be intermittently assessed and interventions provided when indicated.

2. Is the language of the recommendations clear? Does anything need to be clarified or defined further?

Line number (or span)	Comments
4.	- To suggest that prognosis "need not be discussed" at time of diagnosis is impractical and suggestive that it is not a relevant topic. Prognosis is important and does need to be discussed eventually. We are trying to make the point that prognosis discussions are not a requirement at the time of diagnosis, but can be discussed if the patient requests.

27.	Can an example of patients benefiting from MPV be provided for context? We frequently get many requests for MPV, even for bulbar patients. Putting in an example makes this too wordy. The respiratory team at the local centre makes the ultimate selection.
26. 104.	GREAT JOB ON BOTH THIS RECOMMENDATION
105.	In the section for caregivers, “the impact of ALS on patient” can be removed. The entire guideline is for the patient and this section can be dedicated to the caregiver for clarity and emphasis. We would like to keep this phrase in to keep the caregiver involved in planning.

General Comments	
<p>Recommendations are clear and easy to read. Great job!</p> <p>It is a solid guideline for any clinician to access, to provide consistent evidence based support</p>	