ALS Integrated Care Pathway For the Champlain* District

(Amyotrophic Lateral Sclerosis)

March 2008, 2nd Edition

*** Please refer to www@champlain.ccac-ont.ca to learn about the Champlain district in Ontario, Canada.**

disponible en français

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I. INTRODUCTION

i. What is an Integrated Care Pathway?

An integrated care pathway (ICP) is a multidisciplinary outline of anticipated care. This ICP tool presents an overview of the progression of ALS and identifies potential problems that might arise for the client and caregiver as the disease advances. It also offers a list of local resources which can be accessed to support clients with ALS at different stages of the disease. This anticipatory approach to care benefits clients and families by serving as a planning guide for service providers in securing appropriate resources and supports.

ii. Why have an Integrated Care Pathway for ALS?

The goal of the integrated care pathway is to promote and support the development of care partnerships and to provide information to empower clients and their caregivers. Due to the relatively small number of individuals affected with ALS, it is a challenge for agencies with broad service mandates to provide staff with the necessary training and experience to develop a high level of expertise in the management of this disease. This integrated care pathway was developed to assist caregivers in understanding the disease process of ALS from onset to end of life issues.

iii. Who is the Target Audience for the Integrated Care Pathway?

The ICP was developed to assist care providers in the community, both professional and non-professional, but it became clear very early in the development process, that people with ALS and their families would benefit from the ICP as well. Interest from all parties is welcomed as the ICP is seen as a vehicle to educate and inform.

iv. A Message to the Primary Caregiver

When a member of the family is diagnosed with ALS, it can be an overwhelming time. It becomes imperative that a family member, a relative or a friend understand the prognosis of ALS and who to contact during this time of great distress. It is our hope that this document will provide you with a fuller understanding of ALS and offer you the guidance and support you require to empower your decision making.

You are not alone. Most health care partners will come to your aid as needs arise. Nevertheless, as soon as one is diagnosed, families can connect with the ALS Society of Ontario to identify and pursue additional support along the care and support continuum. Taking care of the primary caregiver is often felt to be an indulgence or a secondary issue and, therefore, adequate thought to one's health is often ignored. However, it is again imperative that family caregivers monitor their health with the supervision of the Family Doctor. Service provision and special leave/disability should be considered if the health of the primary caregiver is a risk or might, overtime, become an issue.

Please ensure that this document is made available to your professional care team if they are not already aware of its existence.

v. How should the Integrated Care Pathway be used?

The ICP for ALS has been divided into the following areas:

- Mobility
- Respiratory
- Communication
- Cognitive and Behavioural Changes
- Eating and Swallowing (including oral health)
- Nutrition
- Spirituality
- Sexuality

Each of these sections is then presented according to its stage of progression, based on the client's functional presentation.

The ICP is structured to enable the reader to identify the potential problems or risks which may arise during the course of the disease. It then suggests actions to help manage these problems and local community resources to help support clients and their caregivers. A detailed list of community resources is also available in Appendix A.

While the ICP provides much useful information, it is not intended to address all areas or situations. ***This document is not intended to replace professional clinical assessment and support.***

II. Key Considerations in ALS Disease Management

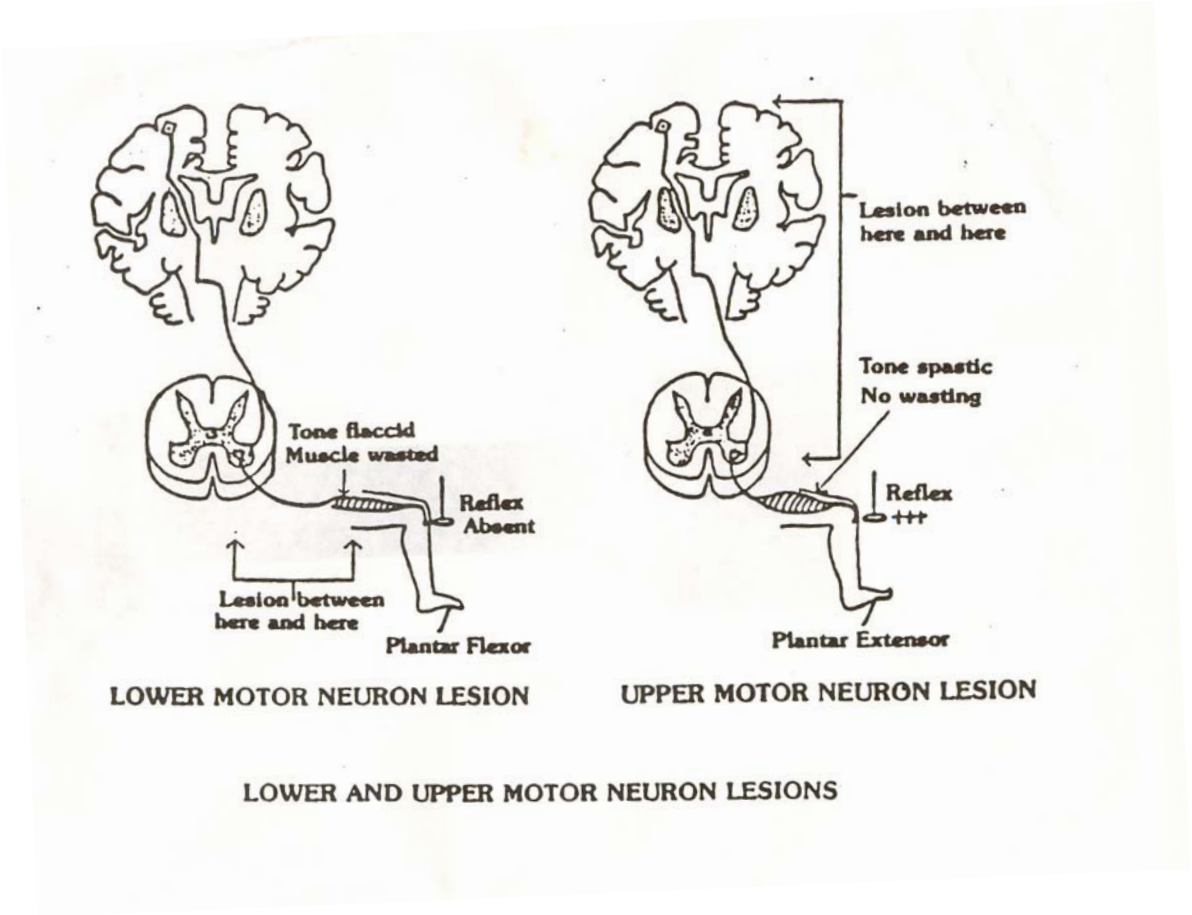
ALS is a complicated and challenging disease. The following 5 overriding messages have been identified as crucial considerations for service providers, clients and families dealing with the illness.

i. Rate of ALS Progression

ALS is a degenerative disease. The rate at which neurons and muscles degenerate is unpredictable and varies greatly from person to person. In some cases the disease process moves slowly, and it may even appear that the client has reached a plateau, while in other cases, it can progress steadily at a rapid rate. Service providers need to be aware of these inconsistencies and be prepared to treat a wide variety of symptoms and an array of functional changes.

ii. Variable Symptomatology

ALS presents itself very differently in different individuals. ALS destroys motor neurons. Symptoms experienced by individuals are largely guided by where the disease attacks. When the motor neurons of the bulbar region of the brain are involved, speaking, swallowing and breathing are affected (Bulbar ALS). When the motor neurons of the spinal cord are affected, one or both arms and/or legs will be affected (Limb Onset ALS). Eventually, as the disease progresses, all areas become involved.



iii. Energy Conservation

Although the course of ALS is unpredictable, fatigue is one outcome that is predictable, resulting from muscle weakness and spasticity. People often complain of tiredness, diminishing strength, and lack of energy. Some of the more noticeable signs of fatigue include slower speech and body movements, lower volume and tone of voice, and shortness of breath. Fatigue can be a very disabling symptom, but by recognizing the signs of fatigue, knowing what exacerbates these symptoms, and learning strategies to conserve energy, the quality of life for individuals with ALS can be improved.

iv. Emotional Impact of ALS

Caring for a loved one with ALS is physically challenging, emotionally overwhelming and financially stressful. The unpredictable rate of disease progression and impaired communication can have a significant impact on personal relationships. Caregivers and family members are at increased risk for depression as well as physical illness and injury related to fatigue and the constant demands of providing care. A care plan should be developed by service providers in conjunction with clients and their families to ensure a holistic approach in managing the ongoing loss of function and socialization related to this terminal illness.

v. Connecting with Knowledgeable Professionals

Access to healthcare professionals who are knowledgeable about ALS and work with the client/family unit as a team is a very important factor in managing the disease process. Members of the interdisciplinary team at the ALS Clinic (The Rehabilitation Centre of The Ottawa Hospital) are local experts for the Champlain District and serve as an excellent resource to clients, families and service providers. It is important for clients to stay connected to their family physician who works in conjunction with the ALS specialist. Family physicians provide ongoing follow up for primary health issues and assessment between clinic visits. It is recommended that all individuals with ALS also be connected with the ALS Society of Ontario (Champlain District) whose mission it is to help people living with ALS and their families manage effects of the disease through such services as the equipment program, as well as support and educational sessions.

Clients will be referred to the Community Care Access Centre (CCAC) as soon as there is a need for home care services. The CCAC will follow the client and family throughout the disease process. Referrals will be made to other services and resources in the community as the need arises. An early introduction to palliative care services can be helpful for some people to cope with this terminal illness. A more complete listing of supportive resources can be found in Appendix A.

As ALS progresses, there are various types of equipment available to assist individuals with mobility, activities of daily living and communication. Choosing the best assistive equipment for their specific situation involves many considerations including an understanding of the rate of disease progression and longer-term needs, as well as issues related to financing and procurement of expensive equipment. ***It is strongly recommended that clients and families consult with a knowledgeable professional before making any major equipment purchases***.

III. MOBILITY PATHWAY

ALS is a disease of the motor neurons in the spinal cord and lower brain which control the voluntary muscles of the body. As these motor neurons die, muscles become progressively weaker and the individual becomes less mobile. The muscles affected, and the order in which they are affected, varies from one person to another. For some people with ALS, the muscles in one or both arms and/or legs are the first to show weakness (limb onset ALS); for others, weakness will begin in the muscles which control speech and swallowing (bulbar onset ALS). As the disease progresses, trunk and breathing muscles become involved.

As muscles weaken, individuals with ALS will typically experience increasing amounts of fatigue, limiting their activities. As body parts become immobile, the associated joints may become stiff, which can be painful. Muscle cramps are also a common symptom of ALS.

It is important to remember that once the motor neurons which supply a given muscle have degenerated, the muscle cannot be strengthened by exercise or by any other means. Stretching and range of motion exercises are often recommended to manage discomfort from joint stiffness or muscle cramps.

Occupational and Physical Therapists will closely monitor the client's fatigue level and abilities over the course of the disease. Equipment, such as small aids to assist with hand function and mobility aids to ease walking and transfers, will be prescribed and updated as the client's needs change. Energy management and safety awareness are key elements in the management of ALS.

ALS Mobility Stages

Stage 1: Mild Motor Symptoms

Presentation
Energy Level
- Still active but tires more easily due to beginning of motor loss

Activities of Daily Living
- Able to manage with normal life and daily routines

Mobility
- May not be able to walk (or shop) as long as they used to and may have some limitations in sports or other physical activities

Psychosocial Considerations:
- Challenges associated with informing family, colleagues and friends about ALS diagnosis
  - Changes to speech and walking due to the neurological condition are often misinterpreted
  - Answering children's questions with age-appropriate information is recommended as they are perceptive and will be aware something is 'not quite right' in the family
- Risk of depression due to reduced energy

Potential Risks	Intervention	Supportive Resources
Reduced energy affecting quality of life	▪ Introduction to energy management ▪ Education re: exercise guidelines (do's and don'ts) ▪ Education re: role of TRC physiotherapist (PT) and occupational therapist (OT) ▪ Education re: partnership between the ALS Clinic and the ALS Society's Equipment Loan Cupboard	ALS Clinic PT, OT, RN and SW ALS Society of Ontario Internet resources: www.als.ca *A Manual for People Living with ALS* (ALS Society of Canada, 2005) Section: Adapting to Changes in Mobility and Maintaining Independence

Stage 2: Moderate Motor Symptoms

Presentation

Energy Level

- Fatigue related to increased effort for certain activities

Activities of Daily Living

- Still independent but requires more time and energy to complete activities

Mobility

- Still independent, but with more difficulty to walk and to get up/down from lower surfaces (e.g. toilet, sofa) due to reduced balance and/or leg/foot weakness

Psychosocial Considerations:

- Reduced social activities
- Reduced intimacy with sexual partners due to fatigue and anxiety
- Financial concerns regarding cost of needed equipment
- Embarrassment and self-image issues associated with need for assistive devices
- Early introduction to palliative care services for support, information and education is beneficial to some clients and their families.

Potential Risks	Intervention	Supportive Resources
Limited energy for activities Injury from falls Pain due to joint stiffness and cramps Potential physical challenges within the client's home environment	▪ Introduction to assistive devices for upper and/or lower body activities (e.g. built up pen for writing) ▪ Equipment prescription to assist with mobility -cane(s), wheeled walker, foot brace (ankle-foot orthosis or AFO) -raised toilet seat -companion chair ▪ Application for disabled person's parking and/or paratranspo permit ▪ Self Range of Motion (ROM) exercises ▪ Self stretching exercises to maintain flexibility and control cramps ▪ Medication for cramping and stiffness ▪ Review home setup to determine adaptability ▪ Referral to community agencies to assist with home maintenance	ALS Clinic PT, OT, Nurse and SW ALS Society of Ontario – equipment loan or shared purchase CCAC OT (as required for in-home assessment) List of vendors for assistive devices (available from ALS Clinic OT and PT) List of contractors to assist with home adaptation (available from ALS Clinic OT) Ministry of Transportation (for accessible parking permit) Resource Material: Handout on Energy Conservation (available from ALS Clinic OT) Community Home Support Agencies and private sector services Internet Resources: The Rehabilitation Centre On-Line Education www.irrd.ca/education ▪ *Energy Conservation* ▪ *Ambulatory aids: a basic guide*

In preparation for Stage 3, it is highly recommended that major equipment purchases be discussed with a knowledgeable health care professional. Since a client with ALS will need more than one piece of mobility equipment at the same time, e.g. a walker and a wheelchair and power wheelchair, it is important to reserve requests for ADP funding for the most sophisticated and expensive equipment. The therapist prescribing the wheelchair has to anticipate equipment needs due to the long wait time in the application process to fund through ADP. Therefore, power mobility and tilt in space have to be prescribed proactively to meet the progressive and often sudden change in physical status.

Stage 3: Advanced Motor Symptoms

Presentation

Energy Level

- Most activities are tiring, requiring frequent rest breaks.
- May nap daily
- Muscle wasting and weight loss
- May have a disabled person's parking permit

Activities of Daily Living

- Requires caregiver assistance for some ADLs
- Increased use of assistive devices to compensate for loss of ability (shower bench, grab bar in bathroom, raised toilet seat, built up utensils)

Mobility

- Walking using furniture, canes or wheeled walker in the home
- Using a wheeled walker or transport/companion chair for outings
- Able to transfer independently from raised surfaces (e.g. raised toilet seat) or needs one-person assist from regular or lower height surfaces (e.g. car, sofa)
- May have difficulty supporting trunk and neck when sitting and/or walking
- May need assistance to maneuver into and out of bed

Psychosocial Considerations:

- Limited social outings
- Client may consider taking one last family vacation before safety issues related to traveling become a concern
- Reduced intimacy with sexual partners due to joint pain, stiffness and fatigue
- Ongoing loss as client faces greater challenges in expressing themselves physically in intimate relationships
- Increased demand on family members to participate in personal care of the individual
- Family may need to consider moving to accommodate the client's (and caregivers) current and future physical limitations

Potential Risks	Intervention	Supportive Resources
Same risks identified in Stage 2, plus: Pain due to poor positioning Client injury due to: ▪ Overexertion ▪ Falls, or ▪ Unsafe assistance technique used by caregiver Caregiver injury from: ▪ Fatigue ▪ Unsafe client assistance technique Reduced ability to do household tasks (laundry, meal preparation) Limited access to places with stairs	▪ Continued education re: energy management ▪ Education on proper positioning for comfort (sitting and lying) ▪ Client and Caregiver education re: safe technique to assist clients (e.g. body mechanics, transfer techniques and equipment) ▪ Equipment prescription for self care, transfers and mobility (built up utensils, transfer board/disc, commode, bedhelper, electric bed, walker, AFO(s), manual/power tilt wheelchair) ▪ Home modifications for accessibility (2nd hand rail for stairs, stairglide, ramps) ▪ CCAC referral for in-home assistance (e.g. PSW) ▪ VHA referral for attendant care ▪ Continue with self ROM and stretching exercises; refer to CCAC for assistance as required ▪ Referral to community agencies to assist with home maintenance ▪ Education re: funding for equipment and services ▪ Education re: potential safety risks when driving ▪ Refer for driving assessment for vehicle adaptation	ALS Clinic OT, PT ,RN, and SW CCAC OT, PT, RN,SW and PSW ALS Society of Ontario Loan Cupboard (equipment loan or shared purchase) VHA Attendant Care Program Community Home Support Agencies and private sector services Refer to community palliative care physician and hospice support program Community building contractors and accessibility funding programs Resource Material: Handout on driving (from ALS Clinic OT) Driving Assessment Program (available through TRC or private companies – a fee is associated with this assessment) Internet Resources: The Rehabilitation Centre On-Line Education www.irrd.ca/education ▪ *Assisted ROM exercises for arms and legs to maintain joint flexibility* ▪ *Principles of transfers for Health Care workers* ▪ See also internet sites recommended in stage 2

Stage 4: Severe Motor Symptoms

Presentation

Energy Level

- Limited physical and social activities due to fatigue
- Regular rest breaks/naps taken one or more times daily
- Continued muscle wasting and weight loss

Activities of Daily Living

- Assistance required for all ADLs with client participating as able
- No longer driving

Mobility

- Using a combination of wheeled walker and manual/power wheelchair for mobility needs (dependent on energy level)
- Bed mobility using electric bed
- May have difficulty supporting trunk and neck when sitting and/or walking
- Transfers with caregiver assistance and equipment (transfer disc, belt and/or board)

Psychosocial Considerations:

- Introduction of electric (hospital) bed increases loss of intimacy (unable to sleep with a partner)
- Lack of privacy in the home due to increased presence of health care providers and equipment
- Increased social isolation
- Decreased self-esteem and self image
- Potential for caregiver burnout (need to consider respite needs of the family)

Potential Risks	**Intervention**	**Supportive Resources**
Same risks identified in Stage 2, plus: Limited endurance for sitting/standing and for walking, due to poor neck and trunk muscle weakness Caregiver fatigue/burnout Social isolation due to complex care needs Skin breakdown due to dependence for mobility and position	▪ Continued education re: safety and energy management ▪ Education on proper positioning; may include prescription of a neck collar ▪ Equipment assessment and update including: o Prescription for wheelchair seating (cushion, tilt, headrest, laptray) for positioning and comfort. o Prescription mattress (pressure relief)	ALS Clinic and CCAC OT/ PT (in collaboration) ALS Society of Ontario Loan Cupboard (equipment loan or shared purchase) CCAC support services (PSW, attendant care, etc.) VHA Attendant Care Program Respite care services and hospice programs

Potential Risks	Intervention	Supportive Resources
	▪ In-home modifications for wheelchair accessibility (porch lift, moving bed to 1st floor) ▪ Assisted ROM (range of motion) exercises and stretches ▪ Reposition in chair/bed to relieve pressure ▪ Regular skin inspection ▪ Request increase hours of in-home assistance ▪ Client should be referred by their family doctor to a physician with palliative care expertise who is able to do home visits	Family Support Groups (Hospice at May Court, TRC, and The ALS Society of Ontario) Palliative care physician Medical Equipment Vendors Assistive Devices Program (ADP) Technical Access Service (TAS)

Stage 5: Loss of Most Motor Control

Presentation

Energy Level

- Very little energy
- Significant weight loss and muscle wasting
- Spends most of the time in the tilt wheelchair or in bed

Activities of Daily Living

- Completely assisted by caregivers

Mobility

- Dependent on caregivers for bed mobility
- Wheelchair dependent (special seating required)
- Transfers using mechanical lift

Psychosocial Considerations:

- Client and family are experiencing issues related to grief and sense of loss
- Client and family may need assistance to plan for celebration of life or funeral arrangements
- Increased financial demands on the family due to equipment requirements

Potential Risks	Intervention	Supportive Resources
Same as Stage 4	▪ Continued education re: safety (patient and caregiver), positioning and energy management ▪ Equipment assessment and update, as required ▪ Assisted ROM exercises and stretches ▪ Increased need for emotional support ▪ Review need for referral to palliative care services (if not already done)	CCAC Social Worker; CCAC OT/PT consults as required CCAC support services (PSW, attendant care etc.) VHA Attendant Care Program Palliative care physician Respite and hospice programs Community of Faith (please see 'How We Can Help" form in Appendix C). Family Support Groups (Hospice at May Court, TRC and The ALS Society of Ontario)

Stage 6 Late stage of illness

Presentation

Energy Level

- Extreme fatigue
- Varying levels of consciousness
- Increased pain/discomfort

Activities of Daily Living

- Complete assistance

Mobility

- Bedridden

Psychosocial Considerations

- Physical contact with the client is very important. Some family members may need to be encouraged to continue to physically connect with the client through avenues such as touching, massage or hugging.

Potential Risks	Intervention	Supportive Resources
Same as Stage 4 and 5	▪ Continued education on safety, positioning and energy management ▪ Assisted ROM exercises and stretches ▪ Increased involvement of visiting and shift nursing ▪ Increased involvement of in-home hospice volunteers ▪ Assessment of spiritual needs and connecting to spiritual resources ▪ Review need for referral to palliative care services (if not already done)	CCAC services VHA Attendant Care Program Respite and hospice programs Spiritual resources Palliative care physician The ALS Society of Ontario

Stage 7 End of Life

Presentation

Energy Level
- Level of consciousness decreasing
- Medications to manage pre-death symptoms

Activities of Daily Living
- Complete assistance

Mobility
- No transfers
- Client is bedridden

Potential Risks	Intervention	Supportive Resources
Increased agitation Difficulty breathing Muscle and joint pain from immobility	▪ Assisted ROM exercises ▪ Palliative care pain and symptom management	CCAC services Respite and hospice programs VHA Attendant Care Program Spiritual resources in the community Palliative care physician Champlain Palliative Pain and Symptom Management Consultation Service

IV. RESPIRATORY PATHWAY

In the early stages of ALS, clients may or may not have started to notice some changes in their breathing especially if they have a sedentary life style. While ALS does not impair the lungs themselves, the muscles involved in breathing and coughing will become weaker over time and eventually result in respiratory failure. In some individuals, respiratory muscles may be affected late in the course of the disease (Limb onset or Non-Bulbar ALS); in others, these muscles are affected early in the disease process (Bulbar ALS).

Early detection of respiratory impairment is essential for timely interventions in planning and implementing appropriate care and reducing the risk of emergency hospitalizations. Common signs of respiratory muscle weakness include inability to take a deep breath especially while lying flat, shortness of breath with activity, general fatigue, morning fatigue, weak cough, excessive mucus and secretions, and low voice volume. Signs and symptoms of advanced breathing impairment include: paleness, bluish color to finger tips, contraction of the neck and other muscles to breathe, confusion and inappropriate sleepiness.

ALS Respiratory Stages

Stage 1: Asymptomatic

Presentation

- May have SOB (shortness of breath) on vigorous exertion

Psychosocial Considerations:

- Loss associated with having to give up previously enjoyed physical activities
- Family sometimes attribute decreased energy as client not trying hard enough to keep up with activities

Potential Risks	Intervention	Supportive Resources
Minimal risks	• Baseline Pulmonary Function Testing (PFT) and Respirology assessment	Pulmonary Assessment Unit at The Rehabilitation Centre (TRC) TRC respiratory therapy staff will provide education sessions on respiratory interventions to any staff caring for TRC / ALS patients (sessions must be at TRC and time permitting) – 613-737-8899 ext 75318 Internet Resources: www.als.ca *A Manual for People Living with ALS* pg. 51-58 (ALS Society of Canada, 2005)

Stage 2: Mild Symptoms

Presentation

- SOB on moderate activity
- May start to have pooling of oral secretions
- Cough is slightly diminished however, client is able to move secretions with some effort
- May have day time fatigue however, no morning headaches

Psychosocial Considerations:

- Increased social isolation due to fatigue
- Altered family relationships as family is involved in performing more caregiving tasks (i.e. lung volume recruitment, assisted cough, suctioning, positioning)
- Decreased self-image and self-esteem due to increased oral secretions
- Early introduction to palliative care services for support, information and education is beneficial to some clients and their families

Potential Risks	Intervention	Supportive Resources
Aspiration of secretions and food Chest infections Hypoventilation	• Allow for rest periods during the day and plan activities to reduce efforts • May benefit from elevating the head of the bed (head and shoulders) • Follow-up visit with respirologist and respiratory therapist o follow-up PFT o introduction of lung volume recruitment with bag (LVR bag) o introduction of LVR with assist cough if indicated o early discussion of advance directives o consider medication to assist with the control of oral secretions • May benefit from oral suctioning with rigid suction tip (Yankauer) • Cold symptoms need to be assessed by the family doctor	Respiratory Protocols for SCI and Neuromuscular Diseases web site http://www.irrd.ca/education/ Lung volume recruitment with resuscitation bag http://www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=7 Manual assisted cough manoeuver http://www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=74

Potential Risks	Intervention	Supportive Resources
Laryngospasm (spasm of the muscles in the throat causing closure of the vocal cords and airway obstruction)	• Avoid factors that may trigger laryngospasm (ie. smoke, strong smells, aspiration, gastric reflux) • Laryngospasm will pass on its own, but may be relieved by dropping chin to chest and swallowing, or by breathing slowly through nose. • Prescription of antispasmodic medication may help with laryngospasm • Reduce the incidence of gastric reflux by raising head of bed and using antacid medication	

Stage 3: Moderate Symptoms

Presentation

- Voice volume is decreased
- May be short of breath at rest or when lying down
- Secretions may be more difficult to control
- Cough is ineffective
- May have elevated carbon dioxide and shallow breathing at night leading to disrupted sleep**,** frequent arousal, daytime headaches and sleepiness

Psychosocial Considerations:

- Impact on energy of family as there is more involvement in monitoring and assisting individual (as well as noise of ventilation equipment and irregular breathing during the night)
- Increased family concern over leaving the individual alone in the home
  - It is important to consider the respite needs of the family to avoid caregiver burnout and negative health effects
- Challenges associated with intimacy and sexual relationships
- Advance directives need to be in place for both client and family caregiver to assist family and caregivers in event of health crisis (Please see Appendix B)
- Increased concern regarding exposure to germs and viruses resulting in increased social isolation and avoidance of crowds
- Individual likely unable to travel by plane due to problems associated with high altitudes
- Client and family may need assistance to plan for celebration of life or funeral arrangements

Potential Risks	Intervention	Supportive Resources

Same as in Stage 2 however, more pronounced
 	
Increasing leaks around the interface when using breathing support systems
 	
Skin breakdown from the interface
 	
Respiratory failure
	• Increase LVR and assisted cough frequency
	• Traditional physiotherapy secretion clearance techniques if LVR is ineffective
	• Elevate head and shoulders if feeling SOB when supine
	• Follow-up visit with respirologist and respiratory therapist
	  ○ follow-up PFT
	  ○ review LVR with bag and assisted cough technique (may need a mask to replace the mouth piece) and necessity for medication/suction to control oral secretions
	  ○ introduction to mechanical inexsufflator if LVR techniques are ineffective
	  ○ discuss advance directives and ventilation choices with Respirologist and life choices with RN and SW
	  ○ ventilation education session with respiratory therapist
	  ○ ventilation trial and initiation of nocturnal ventilation if this is the client's preferred choice, follow-up oximetry and parameter changes when necessary
	• The breathing support interface (mask) may be difficult to adjust. A respiratory therapy vendor in your community can assist the patient in finding the appropriate interface to minimize leaks and optimize comfort.
	• Client should have access to a physician with palliative care expertise who is able to do home visits
		Refer to resource section in Stage 2
		
		Ontario Ventilator Equipment Pool for ventilation equipment questions only
		http://www.vep.ca/
		1-800-633-8977
		
		Mechanical inexsufflator
		  ○ older model http://www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=20
		  ○ new model http://www.irrd.ca/education/slide.asp?RefName=e2r5&slideid=1
		
		CCAC OT (ALS clinic OT) and technician (vendor) to adapt wheelchair to mount ventilator.
		
		TRC, SW and RN

Ventilation = support breathing systems such as a volume ventilator or bilevel (BiPAP)[1] device

Stage 4: Advanced Symptoms

Presentation

- As in Stage 3 however more prevalent
- Using breathing support unit more than 15 hrs per day
- Feeling SOB while on breathing support
- Difficulty with breathing support interface and leaks

Psychosocial Considerations:

- Individual is often unable to leave the home due to an increase in the need for respiratory support and associated equipment (consider portable equipment whenever possible)
- Frustration with difficulties associated with mounting the breathing support unit to an appropriate mobility aid
- Lack of privacy in the home due to increased presence of health care providers and equipment
- Re-visit advance directives to confirm they still reflect the client's wishes

Potential Risks	Intervention	Supportive Resources
As in Stage 3, however more pronounced Difficulty in holding the mouth interface for appropriate ventilation Gastric distention Non invasive ventilation may not be sufficient to support adequate ventilation if bulbar impairment is severe	• Follow-up visit with respirologist and respiratory therapist o follow-up PFT, review LVR techniques o add a back up battery to the breathing support unit, adjust parameters, perform an overnight oximetry and review interface fitting o introduce day time mouth piece ventilation if possible o review medication/suction to control secretions • Increased involvement of visiting and shift nursing ▪ Assessment of spiritual needs and connecting to spiritual resources ▪ Increased involvement of in-home hospice volunteers	Refer to resource sections in Stage 2 and 3 Mouth piece ventilation / LVR with volume ventilator http://www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=51 Respite and hospice programs Spiritual resources in the community Palliative care physician

[1] BiPAP is a trade name for a bilevel ventilator

V. COMMUNICATION PATHWAY

ALS often affects the muscles used in speaking, swallowing, chewing and controlling mucous and saliva. These muscles include the lips, tongue, soft palate, larynx (voice box, throat), and the muscles used in breathing. Weakness and/or poor coordination of some or all of these muscles may lead to difficulty in pronouncing words clearly. Individuals with "Bulbar ALS" experience problems with communication early in the disease progression. The majority of individuals with "Limb Onset ALS" will develop these symptoms as the disease progresses.

Loss of speech is gradual. Speech changes do not occur in the same way or at the same rate for everyone with ALS. This will differ depending on the relative involvement of the upper motor neurons, lower motor neurons and respiratory muscles. It is important to work with a Speech-Language Pathologist (SLP) with experience in ALS and augmentative and alternative communication (anything that supplements or replaces speech) early on. The initial assessment should begin even before there are any speech impairments to ensure speech is monitored over time and teach strategies to help keep natural speech for as long as possible. Alternate means of communication are gradually introduced. Adaptations should be made to ensure the client is able to communicate what they want, when they want. (pg. 45, *A Manual for People living with ALS*, *ALS Society of Canada,* 2005).

ALS Communication Stages

Stage 1: Mild Speech Disturbances

Presentation:

- Mild speech changes with some detectable speech disturbance particularly during times of stress and fatigue. These may include:
  - Some difficulty with articulation and pronunciation (e.g. slurring due to weakness in lips and tongue muscles)
  - Voice hoarseness/strained voice quality
  - Reduced vocal intensity
  - Tongue weakness (may be accompanied by fasciculation or muscle twitching)
  - Nasality of speech with weakening of the soft palate muscles
- Speech remains understandable, but rate, articulation, loudness and resonance may be impaired

Psychosocial Considerations:

- Anxiety and fear due to anticipated communication challenges associated with ALS disease process
- Fear of job loss and loss of self-esteem associated with employment
- Embarrassment due to changes associated with impaired speech
- Challenges associated with informing family, colleagues and friends about ALS diagnosis
  - It is important to inform colleagues about neurological changes that are occurring as they may be misinterpreted (eg. slurred speech)
  - Children will need to be informed as they are perceptive and will be aware something is 'not quite right' in the family

Potential Risks	Intervention	Supportive Resources
Increased difficulty with speech as the day goes on (fatigue)	▪ Provide client the opportunity to ask questions ▪ Provide education re: energy conservation ▪ Provide education regarding compensation strategies to: ▪ minimize environmental adversity (i.e. communicate in a quiet, well lit area, face to face for capacity to read lips) ▪ establish context of message	ALS Clinic SLP Resource Material: *Ways You Can Compensate for Dysarthria* (available from ALS Clinic SLP) Internet Resources: www.als.ca *A Manual for People Living with ALS* (ALS Society of Canada, 2005) ▪ Adapting to Changes in Speech and Maintaining Communication

Stage 2: Mild to Moderate Speech Changes

Presentation:
- Mild to moderate dysarthria (difficulty speaking)
- Increased severity of all symptoms (may include slurring, hoarseness, reduced vocal intensity, nasality)
- Communication becomes more challenging due to increasing fatigue and weakening of the breathing muscles (less air flow available to power the voice)
- Ability to talk in full sentences is diminished
- Client begins to limit complexity and length of messages
- Client will need to use compensatory strategies to change the way he/she speaks to make self understood

Psychosocial Considerations:
- Potential for social isolation due to issues with self-image and self-esteem
- Some individuals are resistant to using communication aids
  - Individuals who are having difficulty communicating verbally may need to be encouraged to use communication aids to reduce the frustration of the individuals and their family/caregivers
- Need to discuss concerns and establish Power of Attorney for Personal Care and Power of Attorney for Property while the individual is able to communicate without significant difficulty (Please see Appendix B)
- Early introduction to palliative care services for support, information and education is beneficial to some clients and their families

Potential Risks	Intervention	Supportive Resources
Client frustration related to altered speech and communication challenges Family and caregiver frustration related to difficulty interpreting client's speech Client's needs not being met by caregiver's inability to understand the client	▪ Early introduction to range of communication options, low and high tech, including AAC (augmentative and alternative communication) ▪ Encourage clients to communicate important messages (letters, last wishes, legal documents) prior to deterioration of speech ▪ Use of compensatory speech strategies by client ▪ Ongoing assessment by Speech-Language Pathologist and adaptation of assistive communication equipment o AAC o Alphabet or word boards (as backups to technology) ▪ May be candidate for oral prosthesis (if soft palate involvement) – palatal lift (prosthedontist)	ALS Clinic SLP CCAC SLP ALS clinic OT adaptation for written communication Technology Access Service Resource Material: *Tips for Understanding Severely Dysarthric Speech* (2004, Pro-Ed Inc.) (available from ALS Clinic SLP) Augmentative Communication and Writing Service (formerly the Technology Access Service)

Stage 3: Incorporation of Augmentative-Alternative Communication

Presentation:
- Moderate to severe dysarthria
- Communication may be limited to one-word responses or highly predictable messages (such as greetings)

Psychosocial Considerations:
- Emotional challenges associated with decreased self-image (including avoidance of sexual relationships)
- Client and family may need assistance to plan for celebration of life or funeral arrangements

Potential Risks	Intervention	Supportive Resources
Increased difficulty using the telephone Increased loss of communication independence	▪ Ongoing assessment by Speech-Language Pathologist for adaptation of assistive communication equipment ▪ Teach strategies for telephone communication Use of 'spokesperson' (family member to assist in translation of client's needs) in situations that require speech	ALS Clinic SLP CCAC SLP

Stage 4: Non-Functional Speech

Presentation:
- Severe to profound dysarthria
- May be able to vocalize for emotional expression or with extreme effort
- Dependent on communication equipment

Psychosocial Considerations:
- Increased social isolation as the disease progresses
- Individual and family may be concerned and anxious regarding financial obligations

Potential Risks	Intervention	Supportive Resources
Inability to communicate with family, friends and caregivers	▪ Develop adequate and consistent 'yes/no' system (which may change rapidly) ▪ Develop eye gaze systems ▪ Enable communication for clients on ventilators ▪ Reassess augmentative communication equipment and strategies ▪ Client should have access to a physician with palliative care expertise who is able to do home visits	ALS Clinic SLP CCAC SLP Augmentative Communication and Writing Service (formerly the Technology Access Service) Respite and hospice programs Spiritual resources in the community

Stage 5: Inability to Speak

Presentation:
- Anarthria (inability to speak)

Psychosocial Considerations:
- Lack of privacy in the home due to increased presence of health care providers and equipment
- Isolation and frustration related to client's inability to make themselves understood or heard
- Family members and caregivers may become frustrated, impatient and emotionally disengaged due to challenges of communication (i.e. time required to communicate)

Potential Risks	Intervention	Supportive Resources
Inability to communicate basic needs to family, friends and caregivers	▪ Potential for use of 'eye gaze' system or other form of yes/no response (providing caregiver is able to discern between yes or no) ▪ Bulbar ALS clients may reach this stage while they have good motor control and may be able to use an AAC device; Others reach this stage at the end-stage of the disease and may not have motor control for AAC usage. At this point, an 'eye gaze' system is the only option. ▪ Increased involvement of visiting and shift nursing ▪ Assessment of spiritual needs and connecting to spiritual resources ▪ Increased involvement of in-home hospice volunteers	ALS Clinic SLP CCAC SLP Augmentative Communication and Writing Service (formerly known as the Technology Access Service) Respite and hospice programs Spiritual resources in the community Family support groups (Hospice at May Court and TRC)

Note. The above information in the Communication Pathway has been adapted from "Management of Speech and Swallowing in Degenerative Diseases, Second Edition" by K.M. Yorkston, R.M. Miller, and E.A. Strand; pp. 19-50, Copyright 2004 by PRO-ED, Inc., Austin, Texas.

VI. COGNITIVE AND BEHAVIOURAL CHANGES

Cognition

Until recently ALS was thought to spare cognitive function. New studies have shown, however, that there is a range of cognitive deficits that occur in the frontal temporal regions due to ALS. The precise incidence has not been established however it is thought that up to 50% of those diagnosed with ALS will have some cognitive changes. There is also a small subgroup (3-5%) that develop a frontotemporal dementia (FTLD) also known as Pick's disease. In these cases the cognitive decline often precedes the physical symptoms.

The cognitive functions that are usually affected include the following:

Insight
Empathy
Reasoning
Initiation
Planning and Organization
Mental rigidity (eg. inability to adjust to a new way of doing things)
New learning

Note: the above information has been adapted from "Cognitive impairment in amyotrophic lateral sclerosis" Lancet Neuronal2007;6:994-1003 by Julie Phukan, Niall P Pender and Orla Hardiman

Potential Risks	Intervention	Supportive Resources
Reduced insight and judgment regarding safety Inability to appreciate caregiver effort (amount of work caregiver is putting in) leading to increased caregiver stress	• Supervision • Cueing to use adaptive aids • Caregiver education around cognitive changes in ALS	• ALS team SW, RN • Referral for private psychotherapy / counseling • Respite services

Involuntary Emotional Expression Disorder (IEED)

Involuntary Emotional Expression Disorder (IEED), also referred to as emotional liability or pseudo-bulbar affect, is the inappropriate and/or uncontrollable laughing or crying that is experienced in some cases of ALS. It is more prevalent in people with Bulbar onset but is also seen in people with limb onset.

Potential Risks	Intervention	Supportive Resources
Social isolation due to embarrassment, misunderstanding by caregivers and exhaustion	• Distraction, such as changing topic of conversation • Discuss use of antidepressant with physician	• ALS team

VII. EATING AND SWALLOWING PATHWAY

With ALS, the muscles in the throat and mouth may be weakened resulting in dysphagia (difficulty chewing or swallowing). Signs of dysphagia include choking on foods or liquids, drooling, increased length of mealtimes, a wet gurgling sound to the voice, coughing and frequent clearing of the throat, and/or weight loss. Unfortunately, in addition to swallowing problems, ALS symptoms also often include weak respiratory function resulting in an inadequate 'protective' cough. Complications can be severe and include aspiration pneumonia (aspiration is when liquids, food, or secretions go down into the airway or lungs and are not removed by effective coughing), dehydration, malnutrition, weight loss and increased muscle wasting due to decreased calorie and protein intake.

Because of the variety of muscles used to chew and swallow, the problems that occur as a result of ALS depend on which muscles have been affected. The dietitian and speech-language pathologist work together with clients to address issues related to the mechanics of eating and drinking (controlling food and swallowing) and alternative dietary solutions in an effort to assist the client to adapt to swallowing problems and maintain good nutrition.

Oral Health

Oral hygiene quickly becomes a challenge for individuals with ALS due to many factors including; weakness in the mouth and throat muscles (which may result in excess saliva and drooling *and*/*or* thick mucous and dry mouth); loss of strength and fatigue resulting in an individual being unable to properly brush their teeth, and increased mouth breathing due to respiratory challenges. Retained food debris, thick mucous, increased saliva and/or dry mouth can all contribute to poor oral health. It is important to maintain good oral hygiene to prevent harmful bacteria from forming and being aspirated into the lungs, as well as to contribute to a positive self-image.

Stage 1 Mild Eating/Swallowing Difficulties

Presentation:

- Complaints of chewing/swallowing difficulties with solid food
- Prolonged mealtimes
- Increased fatigue during mealtimes

Psychosocial Considerations:

- Client may feel socially embarrassed in restaurants or when with guests resulting in increased social isolation
- Increased expectation of caregiver to prepare special meals to adapt texture

Potential Risks	Intervention	Supportive Resources
Decreased nutritional quality of diet Aspiration/Choking	▪ Avoidance of foods that are difficult to chew and swallow (e.g., tough meat, raw vegetables) ▪ Introduction of safe swallowing strategies ▪ Client education on effective coughing technique ▪ Small frequent meals ▪ Continue to maintain good oral hygiene practices including visiting the dentist ▪ Remain focused during eating	ALS Clinic Dietitian, RN and SLP Internet Resources: http://als.ca/als_manuals.aspx ▪ How to Make Eating Safer ▪ Dietary Changes that can Help ▪ Swallowing Problems: Foods to Lose and Foods to Choose

Stage 2: Diet Texture Modifications

Presentation:
- Difficulty managing food and/or liquids
- Progressive weight loss
- Decreased respiratory function contributing to choking and aspiration concerns

Psychosocial Considerations:
- Loss of pleasure of eating food as well as loss of enjoyment of socialization related to shared meals with friends and family
- Choking episodes are very upsetting to family members and friends
- Some caregivers experience feelings of 'guilt' that they can still enjoy food and may choose to not eat in front of their family member or choose a bland diet more similar to what the individual with ALS is eating
- Ongoing risk of social isolation due to inability to eat in public
- Ongoing burden for caregiver to prepare and supervise meals
- Early introduction to palliative care services for support, information and education is beneficial to some clients and their families

Potential Risks	Intervention	Supportive Resources
Choking and aspiration due to dysphagia and increased presence of saliva Dehydration due to inadequate liquid intake Loss of sense of enjoyment typically associated with eating Malnutrition Decreased oral hygiene due to difficulty swallowing and fatigue	▪ Discuss options re: feeding tube (Percutaneous Endoscopic Gastrostomy - PEG tube) – client is more able to tolerate surgery at an early stage of the disease ▪ Modify texture of diet ▪ Swallowing assessment by SLP ▪ Client education on effective coughing technique ▪ Family and caregiver education on assisted coughing technique ▪ Education of family and caregivers re: first aid for choking ▪ Consider using electric toothbrush and flossing aids	ALS Clinic Dietitian, SLP, PT and RN Internet Resources: http://www.irrd.ca/education ▪ Therapy/Treatment - Swallowing Disorders http://als.ca/als_manuals.aspx ▪ Tube Feeding ▪ When to Consider a Tube ▪ Making Your Decision About a Feeding Tube

Stage 3: Moderate Difficulty with Eating/Swallowing

Presentation:
- Increased frequency of choking and aspiration events
- Increased difficulty eating and breathing simultaneously
- More pronounced weight loss

Psychosocial Considerations:
- Clients need emotional support for reactions around insertion of the PEG, including change in body image
- If clients receive the PEG during early stages they may be able to continue to eat/drink orally. This may assist with the acceptance of the PEG
- Caregivers may find the process of assisting with the PEG tube (cleaning, changing) nauseating and have to make an effort to overcome their reactions
- Family and caregivers need to be aware of risk of pneumonia and discuss treatment decisions as part of advance directive in the event of an infection
- Client and family may need assistance to plan for celebration of life or funeral arrangements

Potential Risks	Intervention	Supportive Resources
Choking and aspiration due to dysphagia, fatigue and excess saliva Aspiration pneumonia (risk increases with repeat episodes) Silent aspiration (when foreign material enters the trachea or lungs without an outward sign of coughing or respiratory difficulty by the client) Halitosis (bad breath) Thrush(oral fungal infection) Gingivitis and tooth decay	▪ Insertion of feeding tube if this has been decided by client (i.e. PEG tube – Percutaneous Endoscopic Gastrostomy tube) ▪ Support client's preferred method for receiving nutrition (tube feeding and/or oral feeding) ▪ Adherence to modified texture diet for oral feeding ▪ Use of oral rinses to assist with mouth care (recommended by health care provider) ▪ Suctioning may be necessary to manage excess saliva and/or mucous ▪ Increase liquid intake and use a vaporizer to assist with dry mouth ▪ Discuss medication management of excess saliva with a health care professional ▪ Client should have access to a physician with palliative care expertise who is able to do home visits	ALS Clinic Dietitian, RN, SLP, Physician CCAC SLP and Dietitian Internet Resources http://als.ca/als_manuals.aspx ▪ Mechanics of Swallowing (Be Prepared) http://www.irrd.ca/education ▪ Therapy/Treatment - Swallowing Disorders

Stage 4: Severe difficulty with Oral Feeding

Presentation:
- Severe dysphagia
- Decreased respiratory function (necessary to support eating)

Psychosocial Considerations:
- Cost of specialized formula can be recovered through Ontario Drug Benefit (ODB) for oral/PEG feeding if it is the person's sole source of nutrition (and they are over 65 or being followed by CCAC)
- Travel and visits outside the home are severely restricted unless all feeding equipment can be transported. Transportation of equipment increases stress on caregiver for simple outings over extended hours
- If caregiver goes out alone, substitute care must be found to help around PEG feeding (respite)

Potential Risks	Intervention	Supportive Resources
Airway obstruction/aspiration (all ingestion by mouth is unsafe – however, client may choose to continue with oral feeding regardless of the risk involved)	<u>If client has a feeding tube</u> ▪ Tube feedings as tolerated to maintain hydration and basic nutritional needs <u>If client does not have a feeding tube</u> ▪ Continue to assist client with oral feedings as tolerated	ALS Clinic Dietitian CCAC Dietitian

Note. The above information on Eating and Swallowing has been adapted from "Management of Speech and Swallowing in Degenerative Diseases, Second Edition" by K.M. Yorkston, R.M. Miller, and E.A. Strand; pp.51-65, Copyright 2004 by PRO-ED, Inc., Austin, Texas.

VIII. NUTRITION

ALS and its progression (dysphagia, respiratory function, fatigue) affect nutritional intake. Inversely, malnutrition can affect muscle function, breathing and quality of life.

As yet, there are no recommendations specific to ALS regarding nutritional requirements. Present recommendations remain a nutritionally well-balanced diet e.g. Canada's Food Guide to Healthy Eating.

A person with ALS may face nutritional compromise due to inadequate intake (quality and/or quantity). Clients require an ongoing management plan to minimize risk of aspiration, malnutrition and dehydration while supporting continued enjoyment of food. Despite these strategies, oral feeding may become increasingly slow and difficult and eventually may become a high-risk activity due to inability to meet nutritional requirements and risk of aspiration.

Feeding via percutaneous endoscopic gastrostomy (PEG) has become the intervention of choice when oral intake becomes unsafe or inadequate. However the choice of whether or not to have a PEG remains that of the individual with ALS. The informed choice of the individual should be accepted and supported by caregivers. PEG does not decrease the risk of aspiration. Neither does it artificially extend survival and therefore it is a decision that should be considered separated from tracheostomy and ventilation.

Unlike other aspects of the disease, many of the nutritional issues of ALS do not arise in a sequential order. Thus, they are shown as they might appear.

Decreased Intake of Food/Fluids

Psychosocial Considerations:

- Advance Directives need to be discussed with family and palliative care doctor. Ensure client and family have attended advance directives presentation at TRC to discuss treatment options and palliative care at the end stage of the disease (See Appendix B)
- Families are concerned that the client will suffer if they do not have enough food. Education is required around changes in appetite, desire for food, and changes in the body's ability to use nutrients. Caregivers should be helped in recognizing when feeding their loved one has become more harmful than beneficial

Potential Risks	Intervention	Supportive Resources
Malnutrition due to:		
▪ Decreased appetite due to fatigue and depression	Improve visual presentation of food Introduce small meals and snacks Ensure social interaction at mealtime	ALS Clinic or CCAC Dietitian and RN ALS Clinic or CCAC SLP Internet Resources: http://als.ca/als_manuals.aspx ▪ Adapting to Swallowing Problems and Maintaining Good Nutrition
▪ Problems chewing &/or swallowing food, liquid due to dysphagia, weak lip seal, nasal regurgitation, and presence of thick mucous	Modify food texture of liquid consistency Educate re: safe swallowing strategies	
▪ Decreased pulmonary function resulting in challenges to 'eating/breathing' coordination ▪ Fatigue due to increased time and energy to prepare food and to eat a meal	Small, frequent meals Energy conservation strategies	
▪ Choking due to dysphagia and decreased respiratory function	Training in Heimlich Maneuver and emergency response	
▪ Decreased quantity of food consumed due to decreased appetite, dysphagia and decreased activity	Maintenance of consistent food temperature	
▪ Decreased arm function resulting in decreased ability to prepare meals and decreased ability to self-feed	Provide assistance in meal preparation Introduce adapted tools for eating and drinking	
▪ Decreased quality of intake due to avoidance of problem foods	Nutritional supplements – milkshakes, fruitnogs, commercial supplements	

Decreased intake of fluids

Psychosocial Considerations:

- Advance directives to be discussed with family and palliative care doctor (ensure client and family have attended advance directives presentation at TRC to discuss treatment options and palliative care at the end stage of the disease)

Potential Risks	Intervention	Supportive Resources
Dehydration due to • difficulty swallowing fluids (dysphagia and poor lip seal) • decrease in quantity of fluid intake (due to toileting issues) • increased dependence with toileting	Introduce: ▪ Nectar juices ▪ Thickened fluids ▪ Alternative fluid sources – applesauce, Jell-O ▪ assistance in toileting ▪ alternative equipment, ie. urinal, condom catheter, foley catheter, protective clothing	ALS Clinic Dietician and RN CCAC Dietitian ALS Clinic OT CCAC OT

Elimination

Potential Risks	Intervention	Supportive Resources
Constipation due to ▪ Decreased activity level ▪ Decreased fibre intake ▪ Decreased fluid level ▪ Medications (e.g. amitryptiline, codeine)	▪ Include high fibre foods in the diet (eg. whole grain breads and cereals, vegetables, fruit) ▪ increase fluid intake ▪ modify medications ▪ stool softeners ▪ motility agents ▪ laxatives	ALS Clinic Dietician and RN CCAC Dietitian ALS Clinic OT CCAC OT

IX. SPIRITUAL CARE REFERENCE TOOL

Author: Vivian Stang, M.A. Pastoral Studies, Chaplain, The Rehabilitation Centre, Ottawa, Ontario

ALS is a disease that breaks the connections between body and mind, rendering the body motionless. Often the person with ALS feels estranged from others who are unaware that even in the midst of relentless physical deterioration, the person's mind and heart remain intact.

The Spiritual Care Reference Tool was developed to offer a practical way to enhance spiritual well being and quality of life of persons with ALS. It is designed to identify possible interventions after conducting a spiritual assessment. The tool will help ensure that emotional and spiritual needs are not forgotten or overlooked in the population. The spiritual care described in this tool should be regarded as an essential component of the kind of interdisciplinary, holistic care that can help both persons with ALS and their loved ones move forward into a changed future with a sense of hope and spiritual well-being.

	Early Stage e.g. time of diagnosis	***Intermediate Stage***	***End of Life***
Presenting Emotion	Horror, shock, feeling shattered, denial, anxiety, panic, confusion, uncertainty, worry, sadness, resentment, anger, despair, hopelessness, fear of rejection and/or pity from others, fear of abandonment, isolation, suffering, pain, dying, shame, guilt, demoralization Feeling abandoned or punished by the 'Holy' Repentance, remorse Helplessness, powerlessness, trauma Hope for a cure or that the disease will be retarded by a treatment Gratitude toward others	Sadness and frustration around losses and unanswerable questions Profound grief Hope for relief of discomfort Acceptance Joy of being loved by others	Fear of death Anxiety, uncertainty Relief Contentment Hope for a peaceful end of life
Struggle/Challenge	ALS diagnosis as injury to soul/self/sense of self/self-identity Loss of dignity Existential questioning: Why me? Why am I here? What must I do? What will	Living well with ALS despite multiple and cumulative losses (i.e. autonomy, control over bodily functions, role in family, self-esteem) Living with	Cumulative and final losses Acceptance of terminal stage Realize and accept finitude

	Early Stage e.g. time of diagnosis	***Intermediate Stage***	***End of Life***
	become of me? Am I more than my illness? Grieving for a future one had planned or expected Crisis in relationships (self, others, 'Holy') Renegotiating/redefining intimacy in relationships amidst losses Finding ways to express feelings to loved ones	vulnerabilities and physical suffering Questioning "How will I cope?" and "Can any good come out of my diagnosis/suffering?" Resolving outstanding personal conflicts or tidying personal affairs	
Spiritual Need	Make sense of the senseless ▪ Grapple with the problem of suffering ▪ Find meaning in the diagnosis and meaning and purpose in life/living Find hope, transcend despair Find/maintain a sense of humour, express creativity, appreciate life Recognize the inherent value of relationships ▪ Re-establish relationship to changing self ▪ Maintain/deepen relationships with others ▪ Deepen relationship/connection with 'Holy' Grieve losses Embrace the unknown/mystery in life, live in the present Move from fear to trust Express and accept forgiveness which can lead to spiritual freedom Find a sense of belonging	Move from dependence to interdependence ▪ Help person to bestow gifts as well as receive them ▪ Learn to hold others and be held by others Embrace one's uniqueness, loveableness Grieve losses, practice letting go Maintain a sense of hope, live with courage, bravery, integrity; openness and a growing freedom to be vulnerable Move from disintegration (body and social) to integration (spiritual); move from disconnection (mind-body, message – muscle, thought-speech) to wholeness (in relationships to creation, oneself, others, the 'Holy') Continue to express creativity, focus on pleasure and enjoyment as much as possible	Grieve losses Remember with others Continue to be involved in decision making Deepen awareness of the 'Holy' Bring closure to important issues in life Restore to wholeness/healing even though cure is not possible Say good-bye Final letting go/surrender Live a meaningful death

	Early Stage e.g. time of diagnosis	***Intermediate Stage***	***End of Life***
		Find riches in life despite losses Live life fully, have fun, find blessings in everyday Develop wisdom and insights Move from communication to communion (authenticity and heartfelt communication) Retain a sense of control Embrace the experience vs. resisting one's life	
Possible Intervention	Be sensitive to subjective experience of ALS, process losses and associated grief Encourage expression of and be attentive to emotions (e.g. anger), acknowledge emotional pain and anxieties and facilitate where possible their expression and sharing Allow person to tell their story (many times if necessary); listen for themes in person's story in order to explore meaning of illness Explore core identity with person; assist person in re-establishing new/changing self-identity/sense of self Establish a partnership; provide reassurance that they are not alone Practice noticing the 'Holy' Identify and mobilize a nurturing supportive community Support and respect the	Assist person in 'living the questions' (existential and other) Be attentive to where person is at Reaffirm core identity, remind person that what is important about them is the able, gifted, skilled, capable and full part of themselves; focus on what is possible for the person with ALS Recognize the multitude of strengths a person has Continue to process and respond to grief Refer to clergy Plan for funeral Recommend writing an ethical will, attending an ALS support group, literature reading, writing a letter, doing a life review, leaving a legacy, doing a guided meditation,	Pay attention to issues of abandonment, isolation, pain, discomfort and devaluation of personhood Assess and respond to fears of death and dying Celebrate the life lived Accompany persons to 'the door' Assist person in tying up loose ends, adequately communicate one's love

	Early Stage e.g. time of diagnosis	***Intermediate Stage***	***End of Life***
	person's faith; explore image of and relationship to 'Holy', express anger at the 'Holy' if needed Explore feelings of dread and enhance person's sense of control Explore values/beliefs about life after death Provide encouragement by stating person can still lead a meaningful and high quality of life Encourage couple to continue deepening their intimacy Recommend couples therapy Recommend writing a journal	visualization, practicing ongoing ways to express creativity, using prayer or worship to help cope with illness, practicing mindfulness, relishing the present moment, participating in creative, meaningful rituals (religious or secular), listening to music, heightening a sense of oneness with nature	

X. SEXUALITY

The need for touch, intimacy and closeness is never lost as a result of disability. Sexual expression of love and communication in couples is a very important part of who we are. Amyotrophic Lateral Sclerosis (ALS) does not usually affect a person's ability to have a sexual relationship, including pleasure from tactile sensations, erection, and orgasm, not to mention the pleasure derived from pleasing another person.

Sometimes, however, enjoyment of a sexual relationship may be hampered by symptoms of ALS, such as excessive fatigue, joint pain, arm or leg stiffness or bothersome fasciculation. Some medications prescribed to help with ALS symptoms may also have an effect on sexual functioning.

As the disease causes more weakness throughout their body, people with ALS may worry that they are becoming less attractive to their partners. The partner may refrain from intimacy because they are afraid of hurting the person with ALS during sexual play. It is important to maintain good communication about what works and what does not work for you and your partner.

Many options are available to the individual and/or couple to manage these changes. If you have any questions about how you can maintain enjoyable intimate relationships or if you begin to experience difficulty with some aspect of your sexual well being, please be assured that you can discuss this with any of the health care professionals on the ALS clinical team.

APPENDIX A

KEY SERVICE RESOURCES IN THE CHAMPLAIN DISTRICT FOR ALS MANAGEMENT

A detailed list of available resources and a general description of their role in the continuum of health services is described below.

Resource	Role and Mandate	Contact Information
ALS Clinic The Ottawa Hospital Rehabilitation Centre	The ALS clinic is a multi-disciplinary team consisting of nursing, occupational therapy, physiotherapy, social work, speech language pathology, spiritual care services, clinical nutrition, and physiatry. Also associated with the ALS team are Respirology and respiratory therapists. A patient with their caregiver are seen at the clinic usually every 3 months where they are assessed, provided with recommendations and monitored. Current issues are addressed according to patient's priorities. The team relies on CCAC and other community partners for care delivery and ongoing assessment between visits. The ALS Clinic also provides education sessions and support groups for individuals and their families coping with ALS.	505 Smyth Road Ottawa, Ontario K1H 8M2 (613) 737-7350 ext. 75421
ALS Society of Ontario; Champlain Regional Office	The ALS Society of Ontario provides a range of services to support ALS clients and their families including: ▪ Inservice training for service providers ▪ Information and referral services ▪ Peer support for persons with ALS and their caregivers ▪ Volunteer home visitors In addition, it works closely with health care providers to facilitate the equipment needs of those living with ALS. Through the ALS Equipment Program, the Society provides access to *basic* and *essential* assistive equipment through: ▪ Equipment loaned from our Equipment Pool (shipping cost only) ▪ Equipment Purchase/Rental Assistance (65% client portion) ▪ Communication Equipment Leasing Assistance (65% client portion) This service is available to all persons with ALS residing and/or being treated in the province of Ontario. When health care providers access the ALS Equipment Program the majority of equipment requests can be fulfilled relatively quickly allowing for better utilization of critical funds and timeframes.	1150 Morrison Drive, Suite 204A Ottawa, Ontario K2H 8S9 (613) 820-2267 Toll Free (866) 858-4226 alsont.ca/community/ottawa

Resource	Role and Mandate	Contact Information
Assistive Devices Program	The Ontario Ministry of Health, under the Assistive Devices Program (ADP), helps to pay for a portion of the cost (75%) of equipment and supplies for people who have long term physical disabilities, provided they meet strict eligibility criteria. Devices funded include communication devices and mobility devices (ankle-foot orthoses, walkers/wheelchairs and seating) when an ADP authorizer has prescribed them. Therapists at TRC or CCAC can assess patients for ADP funded equipment. However when a wheelchair is prescribed, a home visit is required by the CCAC OT to allow a functional assessment and identify accessibility issues and solutions to ensure the equipment is compatible with the client's home. ADP only provides funding for one device at a time and expects the ADP authorizer to ensure that the device prescribed will meet the client's present and future needs. The client's own extended health benefits can help pay the balance of funds not covered by ADP. ALS Society's Equipment Program should be considered prior to accessing ADP (please refer to the ALS Society's organization service description for more information)	Ontario Ministry of Health Assistive Devices Branch 7th Floor 5700 Young Street Toronto Ontario M2M 4K5 Tel 1-800-268-6021
Attendant Care Outreach Program (ACOP)	The Attendant Care Outreach Program (ACOP) is funded by the Ontario Ministry of Health and administered by VHA Health and Home Support. This program provides attendant services to individuals with a permanent physical disability who are able to direct their own care. Attendant services are offered on a pre-scheduled basis in the client's home, place of employment or adult educational facility throughout the Ottawa area. Services are available in both official languages, 365 days per year. Attendant Care Services which are non-medical services include such activities as personal care, bowel and bladder management, light housekeeping and laundry etc. Based on the independent living philosophy, attendant care services are flexible and adaptable to client need.	700-250 City Centre Avenue Ottawa, Ontario K1R 6K7 (613) 238-8420 vhaottawa.ca
Augmentative Communication and Writing Service (formerly the Technology Access Services)	The Augmentative Communication and Writing Service is a specialized service of The Rehabilitation Centre that provides Augmentative and Alternative Communication (AAC) services to adults with physical disabilities. AAC refers to a collection of communicative strategies and technologies that support individuals for whom natural speech or writing are not functional. The AAC team is comprised of SLPs, OTs, Rehab Engineers, Electrical Engineers, computer technologists and communication disorder assistants	505 Smyth Road Ottawa, Ontario K1H 8M2 (613) 737-7350

Resource	Role and Mandate	Contact Information
Community Care Access Centre (CCAC)	The CCAC provides in-home health & support services as well as program and service information to the community. Case managers coordinate the delivery of home health care to ALS clients in the community including nursing, personal support, therapy, social work, nutrition and speech language pathology.	4200 Labelle St. Suite 100. Ottawa , Ontario K1J 1J8 (613) 745-5525 champlain.ccac-ont.ca
Friends of Hospice	Friends of Hospice Ottawa is a registered charity dedicated to providing hospice palliative care supportive services to clients in the west end of Ottawa including: In home volunteer support, day hospice program, and bereavement support.	c/o Nepean Community Resource Centre 3730 Richmond Road, Suite 106 Nepean, Ontario K2H 5B9 (613) 838-5744 friendsofhospiceottawa.ca
Home Support Services	The Ottawa Community Support Coalition consists of 19 organizations mandated to provide home-based community support services to seniors and adults with physical disabilities. These agencies deliver, often for a modest fee, a wide range of diverse programs and services including: ▪ Meals on Wheels ▪ Transportation ▪ Home Maintenance and Repair ▪ Friendly Visiting ▪ Security Checks/Reassurance Service ▪ Caregiver Support ▪ Home Help/Homemaking ▪ Foot Care	For more information, contact: Visiting Homemakers Association (VHA) 700-250 City Centre Avenue Ottawa, Ontario K1R 6K7 (613) 238-8420 vhaottawa.ca
Hospice at May Court	The Hospice at May Court is a not for profit community-based organization committed to providing emotional support & practical help for people who are facing a life-threatening illness and their families, regardless of age, religion or culture. The Hospice provides several programs including; home support (volunteer visiting), day hospice and family support programs. The Hospice at May Court also has 9 residential care beds for those in the final weeks of their illness who do not need to be in a hospital setting	114 Cameron Street Ottawa, Ontario K1S 0X1 (613) 260-2906 hospicemaycourt.com
Palliative Care Physician	There are palliative care physicians who specialize in providing palliative care in the home. A medical referral is required. The medical referral is usually made by the client's family doctor but can also be made by the specialist physician treating the ALS.	Accessed through the client's family physician or physician specialist.
Palliative Pain and Symptom Management Consultation Service	Through this service, expert consultants provide support and advice on pain and symptom management to the primary care team caring for the client and the family.	1-800-651-1139
Respite Care	You can discuss options for respite with your CCAC Case Manager and other health care partners. Options for respite may include respite in the home, in a Long-Term Care Home and/or Complex Continuing Care.	Champlain CCAC 4200 Labelle Street, Ste 100 Ottawa, ON K1J 1J8 613-745-5525 champlain.ccac-ont.ca

APPENDIX B

ADVANCE DIRECTIVES AND ALS

It is important that every person has an opportunity to speak with loved ones about what care they would want at the end of life, whether this is medical care offered at the end stage of a chronic illness or as a result of a sudden life threatening trauma.

ALS is a complex illness and many high tech interventions may be offered along the course of the disease. Feeding tubes and assisted breathing devises are often prescribed to enhance quality of life and are not 'end stage' comfort measures. On the other hand ALS patients want to be assured that expert palliative care will be available when they wish to decline invasive medical interventions.

The Rehabilitation Centre at the Ottawa Hospital has developed an advance directive document which is specific to end of life decisions faced by ALS patients and families. This document should also be shared with your family physician and with the palliative care physician (if one is involved in your care). The ALS team will invite patient and families to discuss advance directives and provide detailed information to help them to make these decisions.

As of February 2008 Ontario legislation allows ambulance drivers to provide palliative comfort measures in the home when 911 is called for a medical crisis IF THE PATIENT HAS PREVIOUSLY SIGNED A NUMBERED DO NOT RESUSCITATE FORM. Only your physician or palliative care nurse can provide you with this form and it must bear a specific serial number to be valid for acceptance by emergency service providers. Below is an example of the form. Without this form, if 911 is called, emergency teams will do everything to sustain life and transport the ALS patient to a hospital in a medical crisis. When the patient and family wish palliative care in the home, procedure for comfort measures if breathing or heart stops should be discussed with the physician and CCAC Case Manager.

Ontario Ministry of Health and Long-Term Care

Office of the Fire Marshal

Serial Number _____

Do Not Resuscitate Confirmation Form

To Direct the Practice of Paramedics and Firefighters after February 1, 2008

Confidential when completed

When this form is signed by a physician (M.D.), registered nurse (R.N.), registered nurse in the extended class (R.N. (EC)) or registered practical nurse (R.P.N.), a paramedic or firefighter **will not** initiate basic or advanced cardiopulmonary resuscitation (CPR) (see point #1) and **will** provide necessary comfort measures (see point #2) to the patient named below:

Patient's name – *please print clearly* Surname	Given Name

1. **"Do Not Resuscitate"** means that the paramedic (according to scope of practice) or firefighter (according to skill level) **will not** initiate basic or advanced cardiopulmonary resuscitation (CPR) such as:
   - Chest compression;
   - Defibrillation;
   - Artificial ventilation;
   - Insertion of an oropharyngeal or nasopharyngeal airway;
   - Endotracheal intubation;
   - Transcutaneous pacing;
   - Advanced resuscitation drugs such as, but not limited to, vasopressors, antiarrhythmic agents and opioid antagonists.
2. For the purposes of providing comfort (palliative) care, the paramedic (according to scope of practice) or firefighter (according to skill level) **will** provide interventions or therapies considered necessary to provide comfort or alleviate pain. These include but are not limited to the provision of oropharyngeal suctioning, oxygen, nitroglycerin, salbutamol, glucagon, epinephrine for anaphylaxis, morphine (or other opioid analgesic), ASA or benzodiazepines.

The signature below confirms with respect to the above-named patient, that the following condition (check one ☑) has been met and documented in the patient's health record.

☐ A current plan of treatment exists that reflects the patient's expressed wish when capable, or consent of the substitute decision-maker when the patient is incapable, that CPR not be included in the patient's plan of treatment.

☐ The physician's current opinion is that CPR will almost certainly not benefit the patient and is not part of the plan of treatment, and the physician has discussed this with the capable patient, or the substitute decision-maker when the patient is incapable.

Check one ☑ of the following: ☐ M.D. ☐ R.N. ☐ R.N. (EC) ☐ R.P.N.

Print name in full Surname	Given Name
Signature	Date (yyyy/mm/dd)

- Each form has a unique serial number.
- Use of photocopies is permitted only after this form has been fully completed.

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APPENDIX C

HOW CAN WE HELP A FAMILY CARING FOR SOMEONE WITH ALS
Suggestions for Faith Communities, Neighbours and Extended Family

ALS is an unusual neurological disease which, at the ends stage, may demand many months of intensive caregiving in the patient's home. Homecare and other support programs provide a limited number of hours and family or loved ones are called upon to be available for care, sometimes day and night, while the disease leaves the person with ALS totally dependant for feeding, turning in bed, dressing etc. Friends often ask "how can we help" but the caregivers may be so busy they hardly have time to think how to delegate some of their tasks. Below is a list of task which supportive communities might like to take on. Not all families caring for a person with ALS will need all these tasks but this can serve as a check list which can be reviewed every few months and more support offered as the needs arise.

Master Supervisor(s) to delegate tasks to volunteers _____

NAME OF VOLUNTEER(S)

GENERAL HOUSEHOLD TASKS
Cut grass
Weed garden
Shovel snow
Small household maintenance jobs (specify what needs doing)
Bring in a hot meal (specify times per week)
Do laundry/ironing
Buy groceries 1ce week
Run errands 1ce week
Dusting and cleaning 1ce week
Pick up the mail/ flyers from neighbourhood mail box

PERSONAL CARE FOR PERSON WITH ALS
Take person out for hair cut (or do in home)
In home foot care& manicure
Sit overnight at bedside while caregiver gets sleep
Clean medical supplies in the home
Drive to medical appointments
Drive to church/hospice program or accompany on Paratranspo
Accompany person on leisurely walks (in good weather)
Sit and watch sports/ movies or visit on a scheduled day
Pray or read to them daily
Help person with stretches/body motion exercises/ breathing exercises (exercise instruction would first be taught by a Physiotherapist)

LESS FREQUENT TASKS
Help with accounting/ taxes/banking communication.
Keep records/ filing straight for medical claims etc.
Buying/ adapting clothing (e.g. Knit socks or change buttons)
Set up equipment/ move furniture to improve accessibility in the home.
Be available for Life Line emergency calls In case of falls, etc.

WHEN THERE ARE CHILDREN IN THE ALS HOUSEHOLD
Drive a child to lessons/ sports(specify how often/week)
Help child with homework
Take care of buying a birthday gift and driving child to friend's party
Stay with patient while parent takes child shopping/ or to special events.

B.McIntosh, MSW, RSW. TOH.

January 2008

APPENDIX D

ALS BOOKS AND VIDEOS

The items below (unless otherwise specified) are available at The Rehabilitation Centre Resource Centre. Many of the same titles are also available at the ALS Society Library.

General Information on ALS

- Amyotrophic Lateral Sclerosis: A Guide for Patients and Families – Book
- ALS-It's Not Just Lou Gehrig's Disease – Brochure
- Overview of ALS: A Talk by Dr. Pierre Bourque – Video
- Laugh, I Thought I'd Die: My Life with ALS –Book
- Living and Dying with ALS – Book
- ALS of Canada – Book
- A Booklet for Young People –Book
- Learning to Fall – book
- The Man who Learned to Fall, The story of an ALS Journey- video
- Making Hard Decisions: The Essence of Being Human, Dr Barry Smith – Video
- Tuesdays with Morrie – Book & CD
- Living with ALS (series of videos and manuals available through the Hospice at May Court)

Care Giving

- How Can I Help: A Guide –Book
- Coping With Grief: Strategies for People Living with ALS – Book
- Family Hospice Care - Book
- Caring for Loved Ones at Home - Book
- Caring For The Caregiver - Book
- Self-Care for Caregivers: A Twelve Step Approach - Book
- Self Nurture: Learning to Care for Yourself as Effectively as You Care for Everyone Else - Book
- The Caregiver Series - Coping with Worries - Book
- The Caregiver Series - The Magic of Humour - Book
- The Caregiver Series - Positive Caregiver Attitudes - Book
- The Caregiver Series - Creative Caregiving - Book
- The Caregiver Series - Preventing Caregiver Burnout - Book
- Living Lessons: A Guide for Caregivers – Book
- Easing the Hurt – Book
- Comment puis-je aider – Book
- Faire face au deuil –Book

Dealing with Death

- What Dying People Want –Book
- Facing Death and Finding Hope –Book
- Straight Talk About Death for Teenagers - Book
- Bereaved Children and Teens - Book
- Talking About Death: A Dialogue Between Parent and Child - Book
- When Someone You Love Dies – Book
- The Fall of Freddy the Leaf - Book
- Sad Isn't Bad – Book
- Ottawa doctors talk about risks and benefits of permanent Ventilator (video 1999)

Other topics related to ALS

- Breath of Life – Video
- Easy to Swallow, Easy to Chew – Book

APPENDIX E

ABBREVIATIONS and ACRONYMS

A list of abbreviation and acronyms that are found in this pathway document are found below.

AAC	Augmentative and Alternative Communication
ACOP	Attendant Care Outreach Program
ADL	Activities of Daily Living
ADP	Assistive Devices Program
AFO	Ankle-Foot Orthosis
ALS	Amyotrophic Lateral Sclerosis
CCAC	Community Care Access Centre
ICP	Integrated Care Pathway
LVR	Lung Volume Recruitment
OT	Occupational Therapist (Occupational Therapy)
PEG	Percutaneous Endoscopic Gastrostomy
PFT	Pulmonary Function Test
PSW	Personal Support Worker
PT	Physiotherapist (Physiotherapy)
RN	Registered Nurse
ROM	Range of Motion
SLP	Speech Language Pathologist (Speech Language Pathology)
SOB	Shortness of breath
SW	Social Worker (Social Work)
TRC	The Rehabilitation Centre (of The Ottawa Hospital)
VHA	Visiting Homemakers Association