Living a Quality Life with ALS

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Disclosures

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At the end of this session, participants will be able to:

- Recognize the value of interprofessional palliative care across the continuum of ALS management
- Describe the Canadian Best Practice Recommendations for palliative care and end of life supports in ALS
- Identify strategies to optimize ALS best practices implementation in New Brunswick

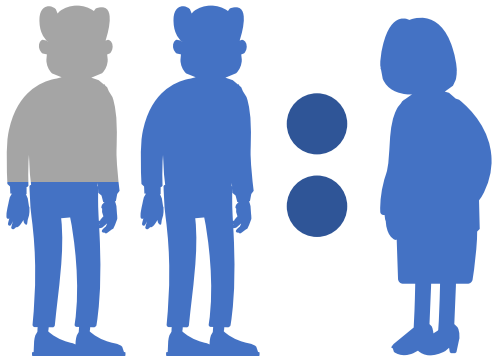
ALS is a progressive, heterogeneous disease

3,000
live with ALS[1,2,4]

1,000
die from ALS each year[4]

2/100,000
new cases of ALS each year[2]

Mean age of onset 55–65 years[2]



1.5 male : 1 female[2]

80% Die within 2–5 years of diagnosis[3]

10% Survive >10 years[3]

1:350 Lifetime risk among men[5]

1:500 Lifetime risk among women[5]

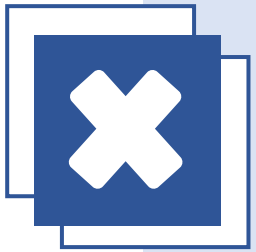
Respiratory complications is the most common cause of death in patients with ALS[6]

While there is no cure, there are treatment options available to improve quality of life, function and survival[1]

1. ALS, MS and MD: How do they differ? ALS Canada. 2020. https://als.ca/wp-content/uploads/2021/05/Fact-Sheet-ALS-MS-and-MD_FINAL.pdf. Accessed December 2021.
2. FYI: Epidemiology of ALS and Suspected Clusters. Jones P (The ALS Association). 2020. https://www.als.org/navigating-als/resources/fyi-epidemiology-als-and-suspected-clusters. Accessed December 2021.
3. ALS Quick Facts. ALS Canada. https://www.als.ca/wp-content/uploads/2017/02/ALS_Quick_Facts-English.pdf. Accessed December 2021.
4. What is ALS. ALS Canada. 2018. https://als.ca/what-is-als/about-als/. Accessed December 2021;
5. Salameh JS et al. *Semin Neurol* 2015; 35:469–76;
6. Niedermeyer S et al. *Chest* 2019; 155:401–8.

The long patient journey to ALS diagnosis

Mean time from symptom onset to ALS diagnosis[1]



13–68% of patients receive an incorrect diagnosis by **PCP or specialist[1]**

Patients visit an average of **3** different physicians **before receiving an ALS diagnosis[3,4]**

Mean time between symptom onset and diagnosis[2]

	Number of Cases	Mean (months)	SD (months)
NS	22	**15.1**	13.1
QC	128	**15.2**	16.4
AB	173	**18.1**	15.5
BC	84	**22.6**	32.2
NB	51	**23.5**	26.5
ON	524	**23.5**	24.9
SK	24	**27.0**	24.4

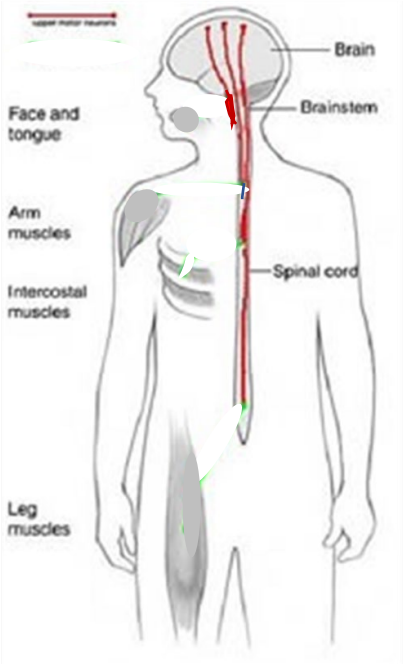
AB, Alberta; BC, British Columbia; NB, New Brunswick; NS, Nova Scotia; ON, Ontario; QC, Québec; SD, standard deviation; SK, Saskatchewan.

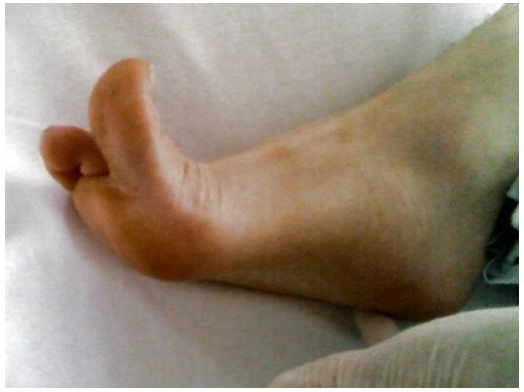
1. Richards D et al. *J Neurol Sci* 2020; 417:117054. doi: 10.1016/j.jns.2020.117054; 2. Hodgkinson VL et al. *Can J Neurol Sci.* 2018;45(6):652–59. 3. Salameh JS et al. *Semin Neurol.* 2015;35(4):469–76; 4. Paganoni S et al. *Amyotroph Lateral Scler Frontotemporal Degener.* 2014;15(5–6):453–6;

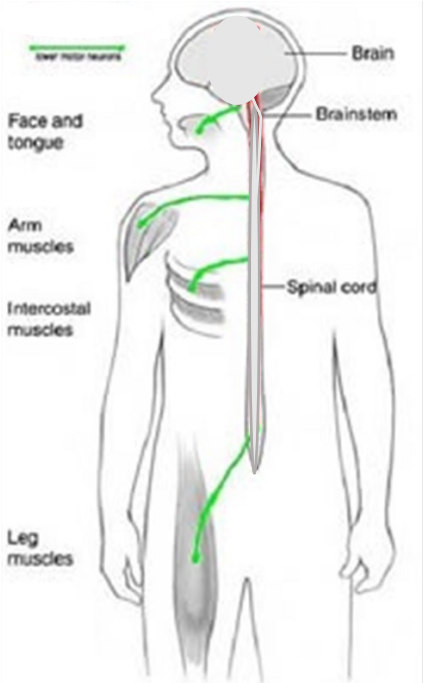
What happens to people with ALS

- Upper motor neuron:
  - Weakness
  - Spasticity, hyper-reflexia
  - Emotional lability
  - Loss dexterity
  - Slowed movement

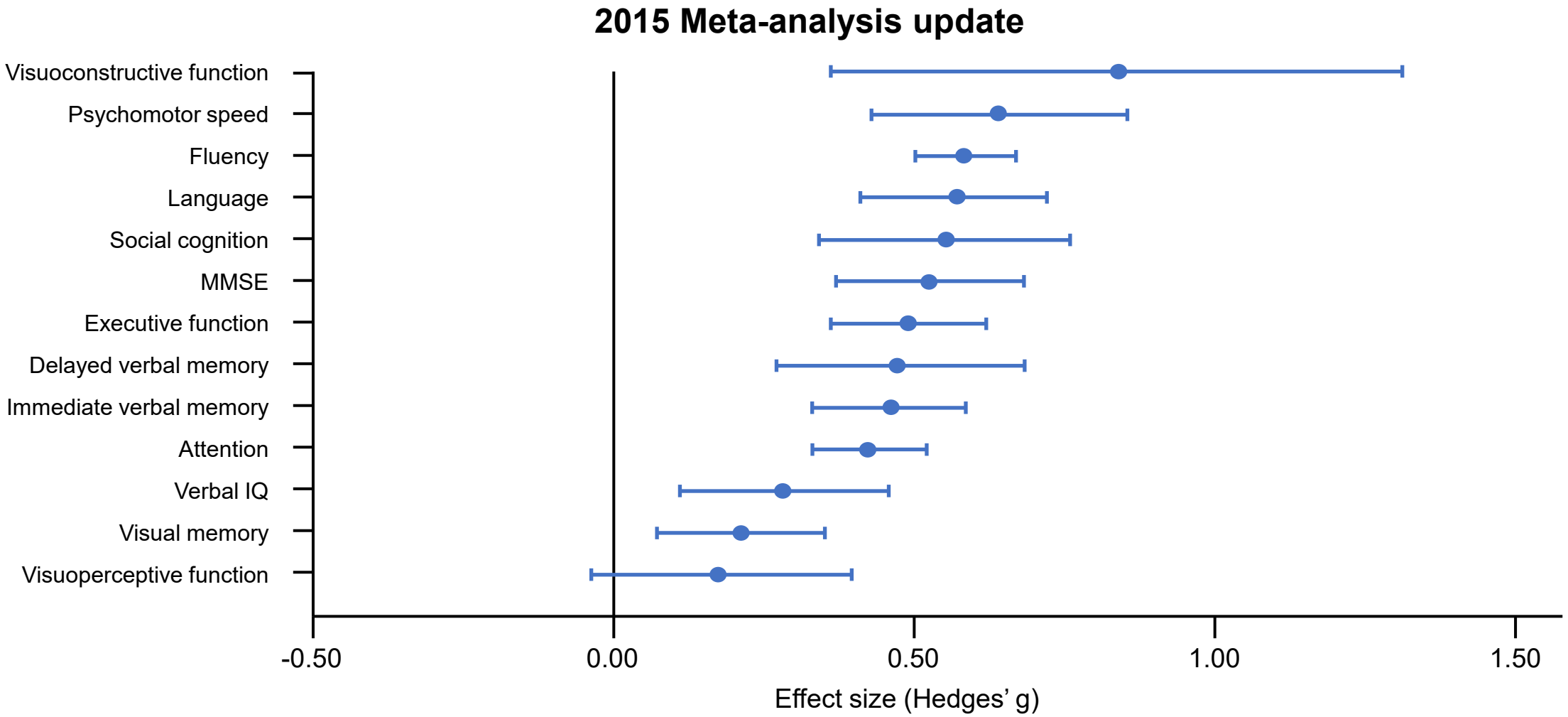
- Lower motor neuron:
  - Atrophy
  - Weakness
  - Hypo-reflexia
  - Muscle cramps
  - Fasciculations







Cognitive impairment



MMSE, mini mental state examination.
Adapted from Beeldman E et al. *J Neurol Neurosurg Psychiatry.* 2016;87(6):611–9.

Presentation of motor neuron disease

Clinical phenotypes

75%	Focal onset of motor symptoms in one limb spreading contiguously over months to become bilateral presentation, usually asymmetrical
25%	Bulbar onset
~50%	Cognitive impairment
15%	Frontotemporal dementia

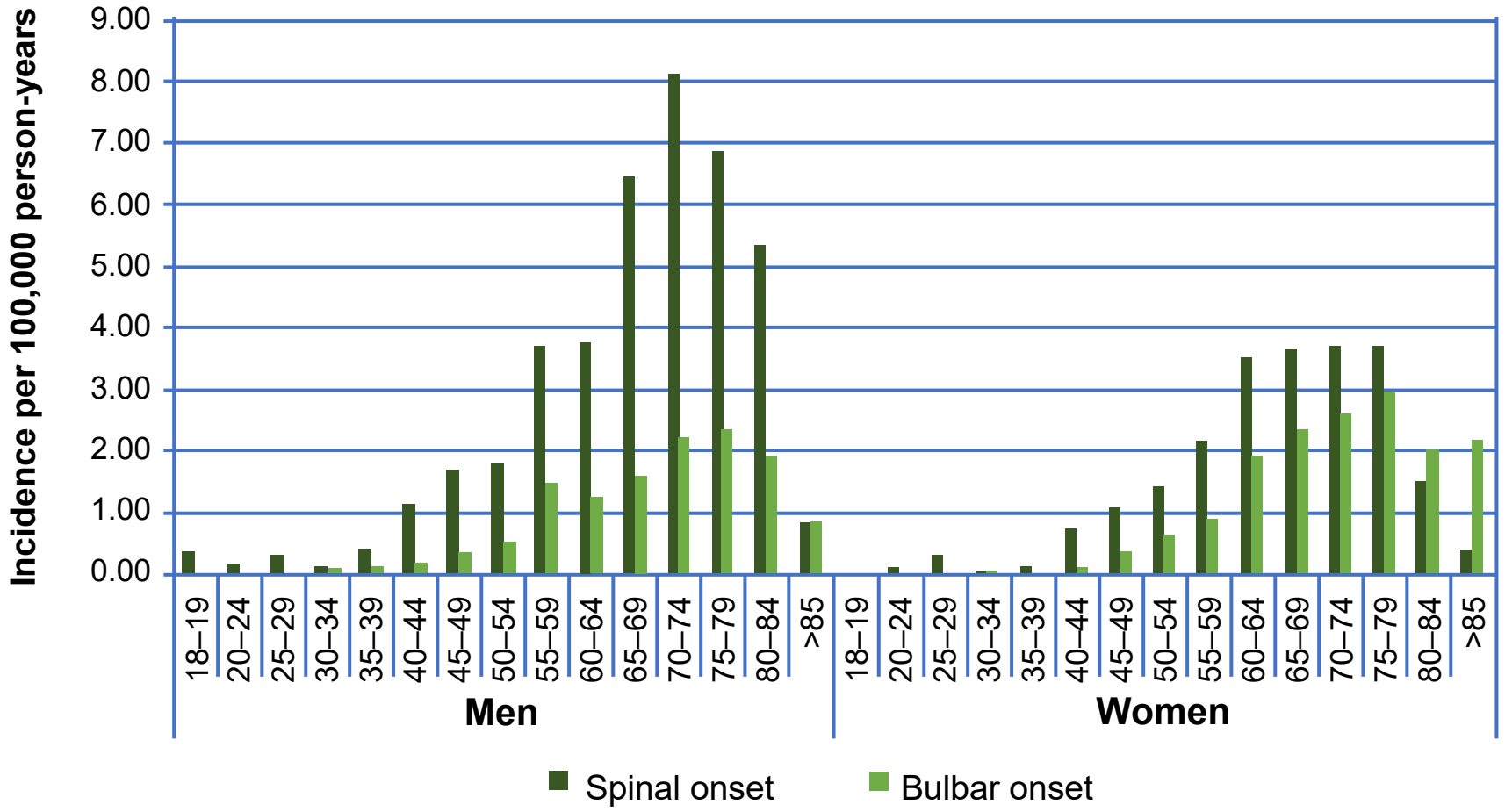
- Primary lateral sclerosis
- Progressive muscular atrophy

Clinical phenotypes based on body region of involvement

- Limb regional variants (e.g., flail limb, man-in-the-barrel syndrome)
- Upper/lower extremity regional variant
- Rare: Mill's syndrome – upper motor neuron – predominant, progressive hemiparesis

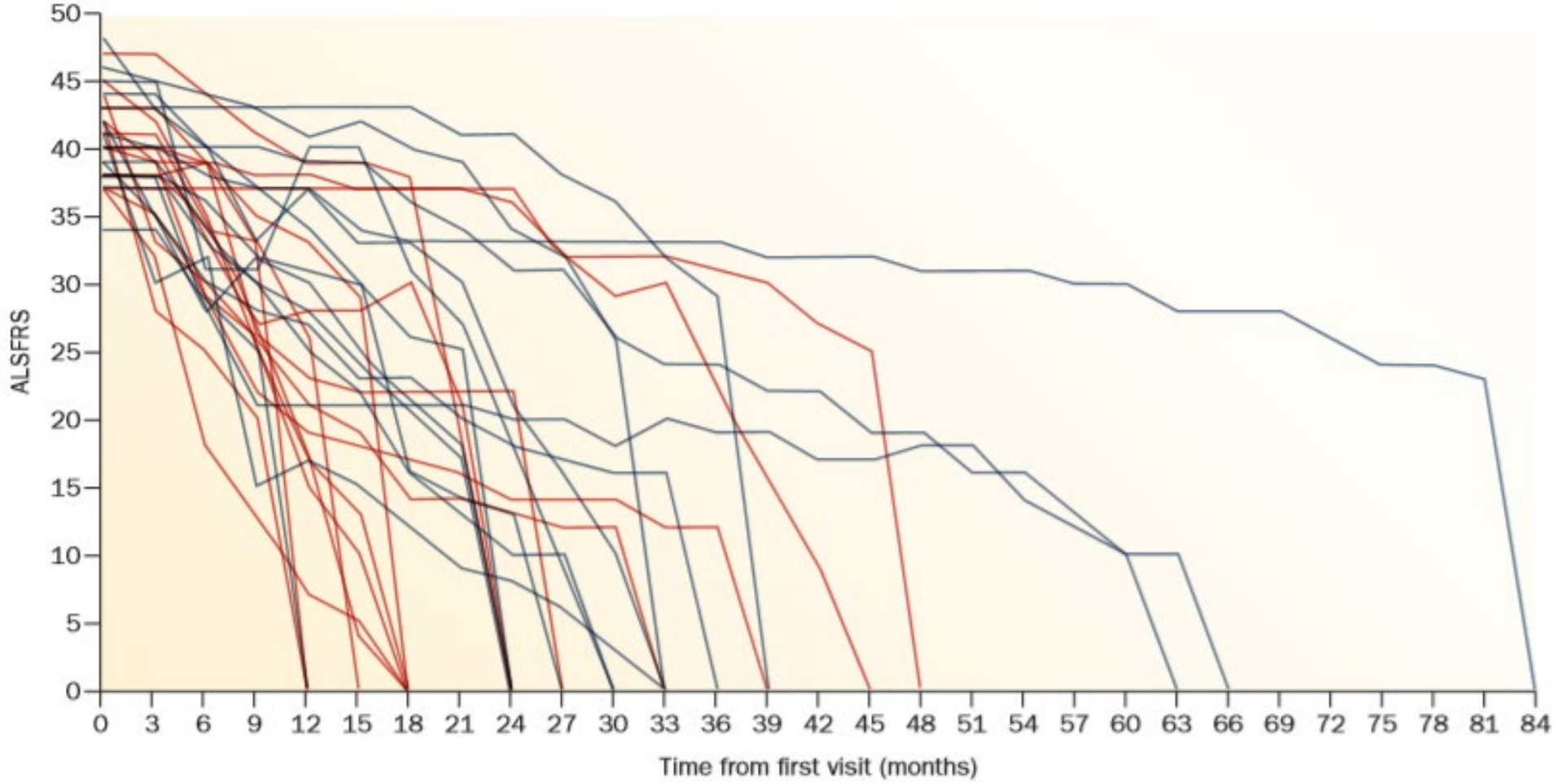
FDA Guidance For Industry. Drug Development for Amyotrophic Lateral Sclerosis. http://www.alsa.org/advocacy/fda/assets/als-drug-development-guidance-for-public-comment-5-2-16.pdf. Accessed May 2019.

ALS can happen at any age



Logroscino G et al. *Curr Geriatr Rep.* 2015;4(2):142–53.

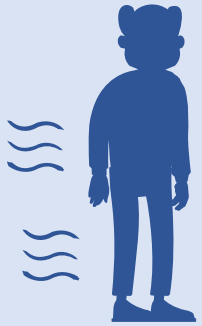
Variability of disease progression in ALS



ALSFRS, ALS Functional Rating Scale.
Swinnen B, Robberecht W. *Nat Rev Neurol*. 2014;10,661–70.

Signs and symptoms for clinical suspicion of ALS

Asymmetrical or progressive, unexplained weakness[1]



Any abnormal EMG with acute and chronic denervation changes[2]

Test results are normal despite progressive symptoms[3]

Elderly person with failure to thrive[4]

Unexplained cognitive and mobility impairment[5]

Combined presentation of upper and lower neurological signs[6]

Pseudobulbar affect[7]

1. Statland JM et al. *Neurol Clin*. 2015;33(4):735–48; 2. Joyce NC, Carter GT. *PM R*. 2013;5(5 Suppl.):S89–95;
3. Bock M et al. *Neurol Clin Pract*. 2017;7(6):488–98; 4. Dziewas R et al. *Dysphagia*. 2017;32(1):78–82;
5. Motor neurone disease: assessment and management. National Institute for Health and Care Excellence.2016. https://www.nice.org.uk/guidance/ng42/resources/motor-neurone-disease-assessment-and-management-pdf-1837449470149. Accessed 22 September 2020; 6. A Guide to ALS Patient Care for Primary Care Physicians. ALS Canada. 2017. https://als.ca/wp-content/uploads/2017/02/A-Guide-to-ALS-Patient-Care-For-Primary-Care-Physicians-English.pdf. Accessed 22 September 2020; 7. Finegan E et al. *Front Neurol*. 2019;10:260.

Dianosis - Gold Coast Criteria

Progressive motor impairment preceded by normal motor function

UMN and LMN signs in >=1 body region OR LMN signs in >=2 body regions

Investigatory studies all excluding other disease possibilities

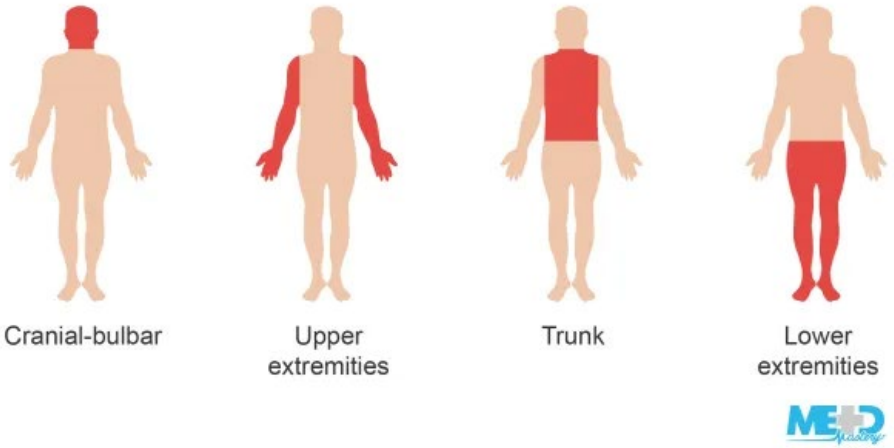
4 Body Regions:

Bulbar

Cervical

Thoracic

Lumbosacral

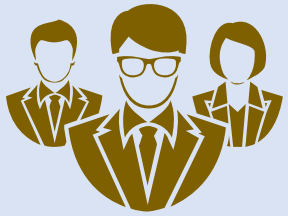


1 Region = 2 limb muscles, different nerves and roots OR 1 bulbar or thoracic muscle

ALS Management – a multi-faceted approach

Disease-modifying treatment

Multidisciplinary care



Symptomatic treatments

Canadian best practice recommendations for the management of ALS

General topics covered

- Communication of diagnosis
- Disease-modifying therapies
- Multidisciplinary care
- Respiratory management
- Nutritional management
- Venous thromboembolism
- Medication alignment
- Symptom management
- Dysarthria
- Exercise
- Cognition and behaviour
- Caregivers
- Palliative care

Shoesmith C et al. *CMAJ* 2020; 192:E1453–68.

Multidisciplinary care

Refer patients to specialized ALS multidisciplinary clinics for optimized health care delivery

Benefits of patient attendance at ALS clinics:

- Increased survival
- Fewer and shorter hospital admissions than patients not attending such clinics
- Increased use of adaptive equipment, riluzole, percutaneous feeding tubes, NIV
- Enhanced quality of life

Frequency of clinical visits should be dictated by **patient's needs** and **rate of progression**

Have a **dedicated nurse** or other **clinic-allied HCP** available to support patients and family members for ALS issues between clinic visits

Multidisciplinary care should take a **team-based approach**, with HCPs addressing issues including:

- Communication
- Nutrition
- Swallowing
- Mobility
- Activities of daily living
- Respiratory care
- Cognition
- Psychosocial issues
- Medical management
- End-of-life care

Telemedicine and **telehealth monitoring** may supplement clinic-based multidisciplinary care

HCP, healthcare practitioner; NIV, noninvasive ventilation.

OT, occupational therapy; RT, respiratory therapy.

PALLIATIVE CARE

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

Palliative care (1/2)

Timing

- **Palliative care services:**
  - Provided by **ALS clinic staff, palliative care practitioners and family physicians** throughout disease course
  - Introduce if severe physical, psychosocial or existential distress
  - Could be introduced before advanced-stage ALS

Withdrawal of ventilatory support

- **Only after consultation/planning** with an HCP with expertise in ventilation withdrawal and palliative sedation
- **Prescribe opioids and benzodiazepines** for anticipatory symptom control **before** withdrawal of ventilation
- **Debrief and offer psychosocial support** for family and HCPs

Treatments

- **Clarify goal of care** with patient → to prolong life or provide comfort-focused care for a good death
- **Assess and manage** factors leading to **breathlessness** (e.g., oral secretions & anxiety)
  - Consider **opioid titration** and **air flow** across the face
- **Introduce ACP** early/when patient inquires and discuss routinely during follow-ups
- Encourage patients to discuss **preferences for end-of-life care** with family and caregivers
- **Integrate palliative care** into routine management before terminal phase
- **Continue NIV and PEG tubes** for symptom relief and quality of life, as per patient preference

ACP, advanced care planning; HCP, health care practitioner; NIV, noninvasive ventilation; PEG, percutaneous endoscopy gastrostomy.

Palliative care (2/2)

Bereavement

- Provide **psychosocial support** for bereaved caregivers
- Initiate early discussion and support for bereavement process, even before patient's death

Organ donation – patients with ALS

- **Solid organ donations** may be accepted, as determined by local organ donation organizations
- **Tissue donations** are not accepted (e.g., corneas, skin or bone); may be able to donate for ALS research
- Clinics should **direct donation inquiries** to provincial organ donation organizations

Medical assistance in dying (MAiD)

- Direct discussions about MAiD to a physician or nurse practitioner, abiding by regional guidelines
- Physicians caring for patients with ALS **must** provide access to information about MAiD when requested
- **Do not assume** questions about MAiD constitute a request for MAiD – Allow this to open a discussion about end-of-life care and ACP
- Provide concurrent palliative and supportive care for patients pursuing MAiD

Symptom management (1/3)

Pain

- Is a recognized consequence of ALS
- **Regularly assess** and tailor treatments to specific cause

Fasciculations

- Often does not require medication management
- Consider **gabapentin** if fasciculations cause substantial distress

Sialorrhea

- First-line therapy: **anticholinergic medications** (consider switching if one is ineffective)
- **Oral suction** as an adjunct therapy
- Second-line therapy: **botulinum toxin** (consider *after* feeding tube insertion)
- Second- or third-line therapy: **salivary gland radiation**

Pseudobulbar affect

- May not be a symptom of depression or impaired cognition; **educate patients** and families
- No treatment required unless patient is distressed
- Consider medications that co-treat **concomitant symptoms** (e.g., SSRI for depression, etc.)
- Consider **dextromethorphan** (20 mg) with **quinidine** (10 mg)

SSRI, selective serotonin reuptake inhibitor.

Symptom management (2/3)

Spasticity

- **Stretching** can help
- Consider **baclofen, tizanidine, botulinum toxin, benzodiazepines and cannabinoids**
- Insufficient evidence for intrathecal baclofen

Cramps

- Differentiate muscle cramps from other causes of pain
- **First-line therapy:** tonic water, gabapentin, baclofen
- **Second-line therapy:** quinine, levetiracetam, mexiletine

Depression

- Consider **SSRIs** or **SNRIs**
- Consider **nonpharmacologic supports** (e.g., psychology, social work, psychiatry or spiritual care)

SNRI, serotonin-norepinephrine reuptake inhibitor; SSRI, selective serotonin reuptake inhibitor.

Symptom management (3/3)

Anxiety

- Determine if related to **respiratory insufficiency** – if present, treat appropriately
- Prescribe an **SSRI** if depression is also present
- Benzodiazepines can exacerbate respiratory insufficiency
- Consider **nonpharmacologic supports** (e.g., psychology, social work, psychiatry or spiritual care)

Insomnia

- **Investigate cause**; i.e., respiratory insufficiency and depression
- Consider **respiratory investigations and sleep studies** to determine type/cause of insomnia
- **Pharmacologic management** depends on cause

Fatigue

- Consider **reversible causes** (e.g., respiratory insufficiency, sleep disorders, depression, medication AEs and riluzole use)
- If taking **riluzole**, consider reducing or discontinuing
- Consider an **occupational therapist** to discuss energy conservation techniques

AE, adverse effect; SSRI, selective serotonin reuptake inhibitor.

Dysarthria

Patients with dysarthria should be regularly followed by a speech language pathologist to ensure timely communication interventions

Offer augmentative and alternative **communication devices** and strategies to eligible patients
– These may also **reduce caregiver stress**

Access to different modes of communication, including **social media**, can allow independence, participation and **better quality of life**

Tailor communication devices **to needs** and abilities

- Patients with cognitive impairment may need **individualized** strategies
- Offer **voice amplification** for reduced vocal projection
- Offer **voice banking** to appropriate patients

“Positive Tiredness”

- Being tired isn’t always a bad sign!
- The feeling of being tired (particularly post-exercise) needs to be considered normal
- Encourage ALS clients to participate in mildly to moderately challenging exercise
- Energy Conservation Strategies- management of “tiredness” or “fatigue”

Don’t Overdo It:
Energy Conservation Strategies

PRIORITIZATION

PLAN

PACE

POSITIONING

Don’t Overdo It:
Energy Conservation Strategies

DO

participate in the activities that are important to you and that you can navigate safely

DELAY

tasks until tomorrow or later in the week when you don’t have as much on your plate or have more energy. Not everything has to be done in a day!

DELEGATE

tasks - ask others for help

DUMP

just don’t do it!

Cognition and behaviour

Perform screening for cognitive and behavioural impairment early

For concern with cognition or behaviour, **arrange specific assessments** for patient and family members/caregiver, as appropriate

No studies on use of **pharmacologic agents** to manage cognitive/behavioural impairment

Cognitive or behavioural impairment should not preclude recommendations for NIV or gastrostomy insertion

Discuss **challenges of intervention compliance** with cognitive or behavioural impairment with patient and family **before** proceeding with an intervention

Frontotemporal dementia negatively affects survival – do **ACP early**

Consider a **multidisciplinary approach** to manage problematic behaviours

Consider involving a **behavioural specialist** (e.g., occupational therapist or psychologist) or **psychiatrist**

ACP, advanced care planning; NIV, noninvasive ventilation.

Caregivers

**Be attentive to the needs and emotional well-being of caregivers –
Involve caregivers in planning**

Multidisciplinary clinics should be aware of the **financial strain** on caregivers – Provide information for **relief programs,** where possible

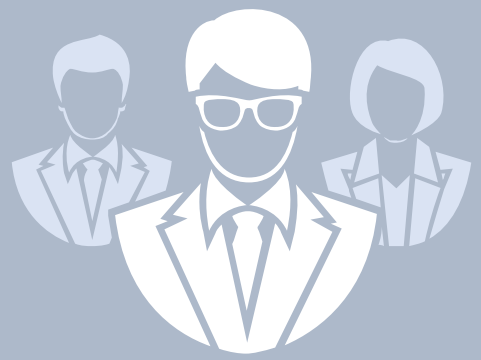
Assess **caregiver burden**, **coping strategies**, **mood** and **family dynamics** to help identify caregivers and families in need of respite and support services

Local ALS societies may have **resources** for family members and caregivers

Best practice recommendations

Key takeaways

Management of ALS patients requires specialized multidisciplinary care



1

Respiratory, nutritional and symptom management should factor patient preferences, including when to stop interventions

2

Caregiver support, palliative care and advance care planning are important components of disease management

3

Implementation

Strategies to optimize ALS best practices implementation

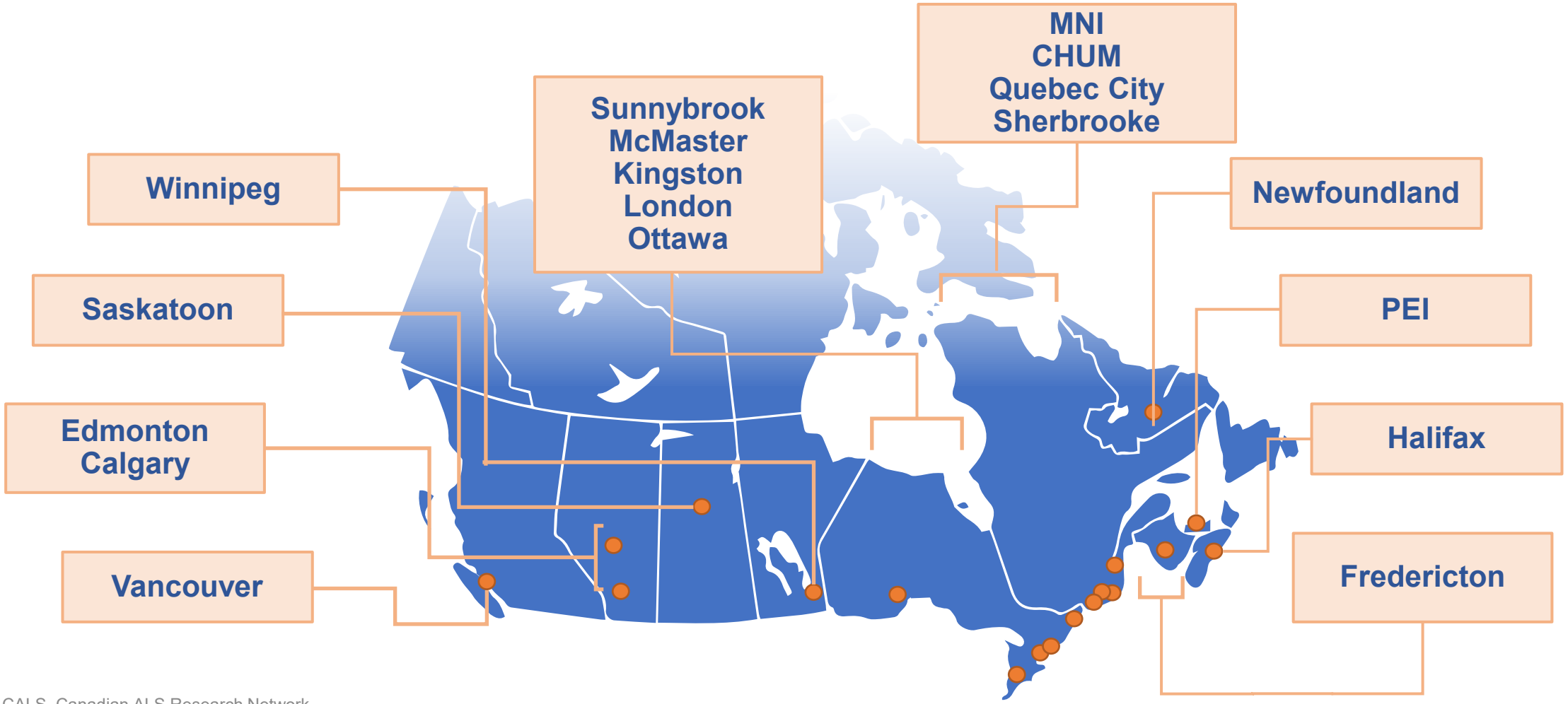
Canadian ALS Research Network (CALS)

- National network of clinicians/ALS research and care clinics
- Established in 2008
- Mandate:
  - Attract and conduct multicentre, ALS clinical research studies
  - Participate in international collaborations with other ALS research consortia
  - Promote education initiatives

Since the formation of CALS, the number of Canadian patients participating in ALS clinical trials has increased dramatically

Zinman L et al. *Clin Investig (Lond)* 2013;3(12):1113–17.

CALS members include researchers and staff from 18 ALS research centres in Canada



CALS, Canadian ALS Research Network.

SIGNS/SYMPTOMS & CLINICAL FEATURES

Amyotrophic lateral sclerosis (ALS) is a heterogeneous disease that can be difficult to diagnose. It is critical to identify both the **onset AND progression** of these symptoms:

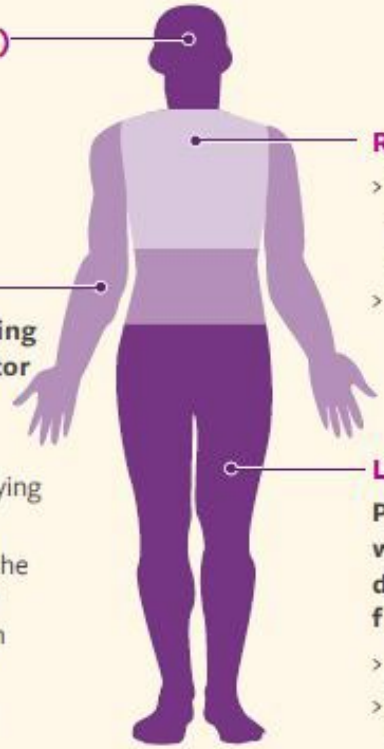
HEAD AND NECK (BULBAR)

- Emotional lability
- Slurred speech
- Difficulty swallowing
- Excess saliva

UPPER BODY

Progressive weakness resulting in asymmetric decline in motor function:

- Impaired handwriting
- Difficulty lifting, reaching, carrying
- Difficulty with everyday tasks (e.g., preparing food, starting the car, using keys, opening jars or bottles, retrieving change from pockets, etc.)
- Trouble with dressing/hygiene (e.g., doing buttons, cutting fingernails, etc.)



RESPIRATORY

- Shortness of breath with exertion or activities of daily living
- Orthopnea

LOWER BODY

Progressive asymmetric weakness resulting in a decline in gross motor function:

- Frequent tripping
- Difficulty on stairs, getting out of a chair, standing on toes, etc.
- Foot drags when walking; cannot walk as long/far

REFER IMMEDIATELY

- ☐ **Asymmetry + Progression** (progression of the motor syndrome within a region or to other regions)
- ☐ **Progressive Speech and/or Swallowing Difficulties**
- ☐ **Pseudobulbar Affect**

CONSIDER A REFERRAL

- ☐ Cognitive complaints/symptoms in presence of mobility impairment/weakness
- ☐ Upper/lower motor neuron signs
- ☐ Failure to thrive in elderly patient
- ☐ Patient suspects ALS

NOTE THESE COMMON ALS MISDIAGNOSES

- **Carpal Tunnel/Sciatica/Ulnar Neuropathy** are UNLIKELY if patient exhibits hand weakness/ foot drop with no pain or sensory loss
- **MSA, Parkinson's, Alzheimer's** may have SIMILAR symptoms

ALS SLA CANADA

CALS Canadian ALS Research Network

The ALS Society of Canada has worked closely with the CALS Network and funding partners to present this critical referral information to you. For more information visit: als.ca/referals or contact referALS@als.ca

DON'T DELAY, REFER ALS. RIGHT AWAY.

WHEN TO REFER?

Referral to a specialized Canadian ALS Research Network (CALS) Clinic should occur...

...AS SOON AS ALS IS SUSPECTED

...PRIOR TO COMPLETION OF DIAGNOSTIC TESTING

...IN ABSENCE OF A DEFINITE DIAGNOSIS

HOW TO REFER?

1. ORDER TESTS AND REFER

Refer to your nearest CALS Clinic, while these results are pending:

- ☐ **Pulmonary Function Test (FVC)**
- ☐ **Electromyography (EMG)**
- ☐ **Imaging**

IT IS ALSO BENEFICIAL TO:

› Indicate if your patient is aware they are being referred to a CALS Clinic

› Consider genetic testing when there is an indication of family history

QUESTIONS? REFERALS@ALS.CA

2. COMMUNICATE THE REFERRAL TO YOUR PATIENT

Share and Review the Patient Tool (page 3) with your patient.
Discussions on prognosis DO NOT need to take place during initial diagnosis/ referral stages unless specifically requested.

YOU MAY FIND IT HELPFUL TO SAY:

- *"I'm concerned that there are signs that this could be a serious neurologic disease. I want to refer you to another expert — a neurologist or physiatrist — who will see you in about 2-6 weeks to assess/re-assess your symptoms."*
- *"This is not a diagnosis yet. The CALS clinic is best equipped to do further testing and may be able to provide you with better care supports for your symptoms."*

QUESTIONS ABOUT YOUR SYMPTOMS?

If you have concerns about any of these symptoms, please bring them to your CALS Clinic appointment:

- ☐ TROUBLE BREATHING
- ☐ SPEAKING PROBLEMS
- ☐ EXCESS SALIVA
- ☐ TROUBLE SWALLOWING
- ☐ EXCESSIVE LAUGHING & CRYING
- ☐ PAIN
- ☐ MUSCLE TWITCHES
- ☐ TIGHT LIMBS
- ☐ CRAMPS

Your CALS Clinicians may also discuss these symptoms and other topics with you:

- ☐ DEPRESSION
- ☐ ANXIETY
- ☐ INSOMNIA (TROUBLE SLEEPING)
- ☐ FATIGUE
- ☐ NUTRITION
- ☐ EXERCISE
- ☐ COGNITION (THINKING) and BEHAVIOUR

Any or all of these tests may be used to help diagnose ALS:

- Blood and urine studies
- Breathing tests
- Magnetic Resonance Imaging (MRI)
- Muscle and nerve function tests

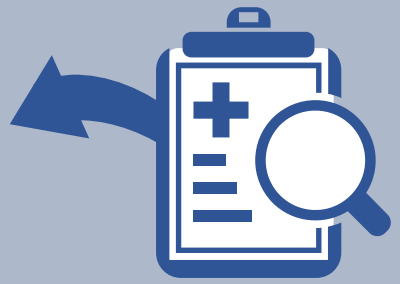
ALS Society of Canada has worked closely with the CALS network and funding partners to present this important information to you. For more information visit: als.ca/research/canadian-als-research-network or contact referALS@als.ca.

ALS SLA CANADA

CALS Canadian ALS Research Network

Patients can be referred to a specialized CALS clinic

As soon as ALS is suspected



Prior to diagnostic testing being completed

In absence of a definite diagnosis

CALS, Canadian ALS Research Network.

CANADIAN BEST PRACTICE RECOMMENDATIONS
FOR THE MANAGEMENT OF AMYOTROPHIC LATERAL SCLEROSIS

ALS SLA CANADA

To enable ALS clinicians, allied health professionals and primary care providers to align clinical care with best practices to ensure people living with ALS receive consistent care no matter where they live in Canada.

130+
Recommendations across 13 Areas

ADDRESS ISSUES RELEVANT TO THE CANADIAN CONTEXT

- Timeliness of care
- Treatments
- Caregiver Support
- Medical Assistance in Dying (MAiD)

MULTIDISCIPLINARY CARE FOCUS

a specialized approach with attention to the whole person, including their emotional aspects of wellbeing

Empowers **INFORMED, COLLABORATIVE DECISIONS** and advocacy for optimal care

FIRST

comprehensive resource to inform ALS care in Canada

ICE BUCKET CHALLENGE

donations put to work

Developed by a 13-member **PAN-CANADIAN** working group

3,000

In Canada, about 3,000 people live with ALS, a progressive, degenerative disease that gradually results in paralysis.
With no cure, 4 out of 5 people with ALS will die within 5 years of being diagnosed.

TO LEARN MORE, VISIT **ALS.CA**

Shoesmith C, Abrahao A, Benstead T, et al. Canadian best practice recommendations for the management of amyotrophic lateral sclerosis. 2020 November 16;192:E1453-68. doi: 10.1503/cmaj.191721

ALS SLA
CANADA

Margot Algie, living with ALS since 2015

CANADIAN BEST PRACTICE RECOMMENDATIONS
FOR THE MANAGEMENT OF AMYOTROPHIC LATERAL SCLEROSIS

ESTABLISHING A NATIONAL STANDARD FOR ALS CARE AND TREATMENT IN CANADA

AN OVERVIEW

February 2022

SYMPTOM MANAGEMENT

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

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

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

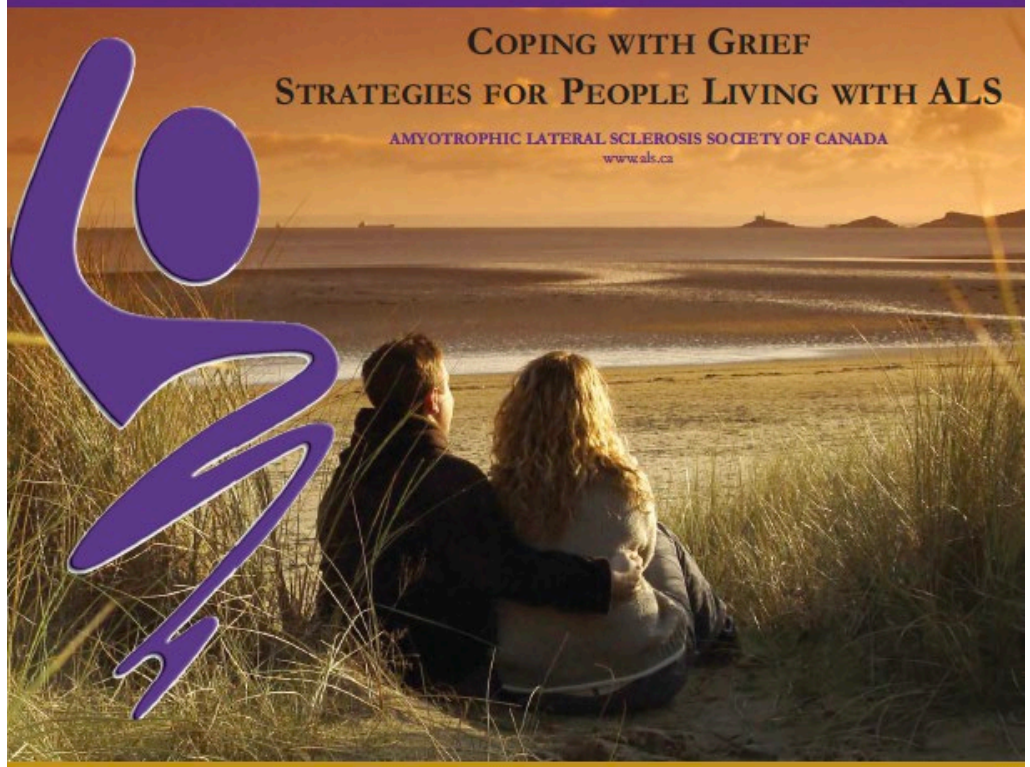
PAIN

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

MUSCLE TWITCHES (*fasciculations*)

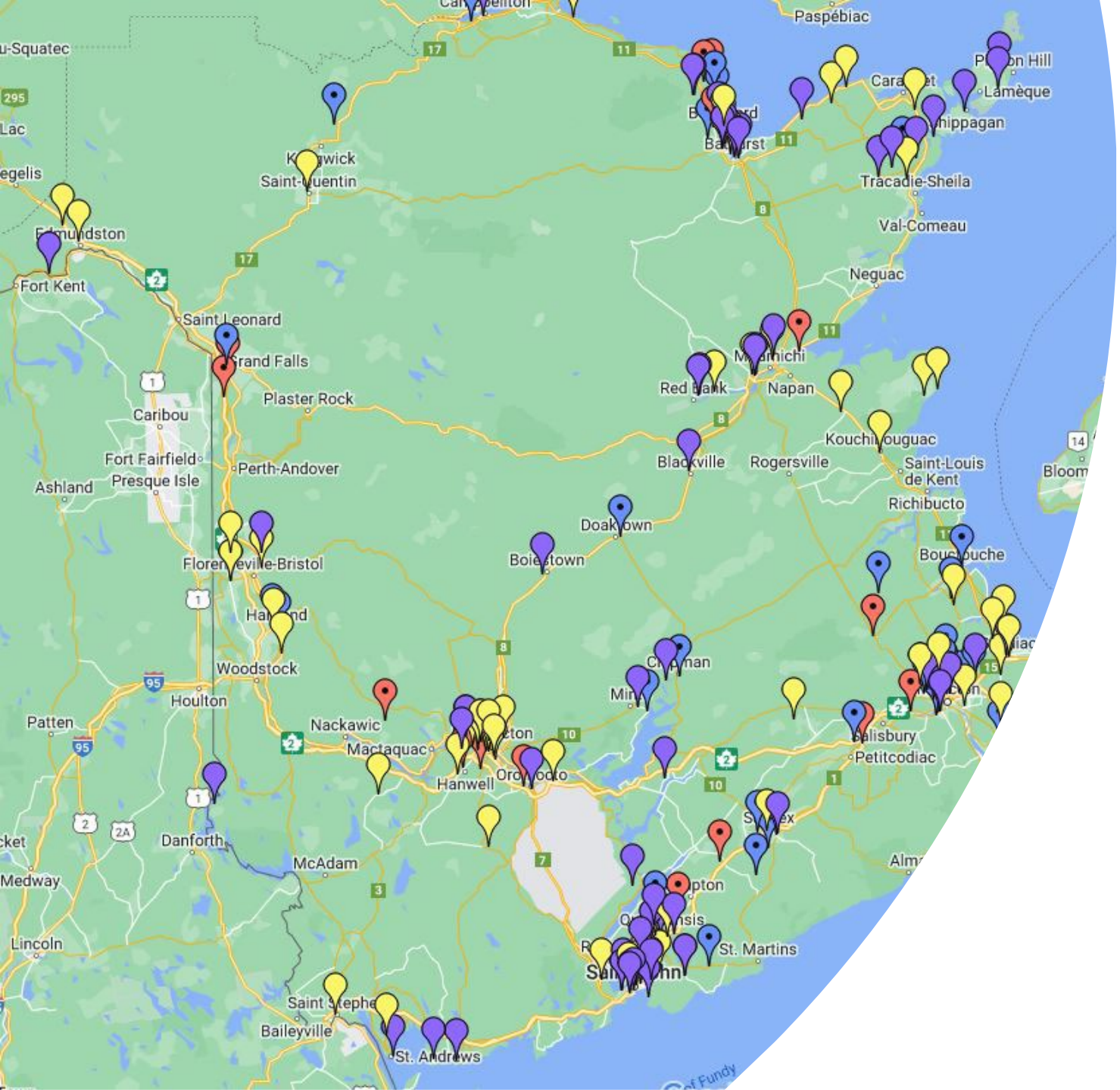
- In most people living with ALS, fasciculations do not need to be managed with medication.
- If they cause substantial distress, the medication gabapentin can be considered.

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



Coping with Grief
Strategies for People Living with ALS

AMYOTROPHIC LATERAL SCLEROSIS SOCIETY OF CANADA
www.als.ca



Where do you live?

Travel and weather

Accelerated Implementation

ELSEVIER

Telerehabilitation

Principles and Practice

2022, Pages 135-148

Chapter 10 - Telerehabilitation in Amyotrophic Lateral Sclerosis

Colleen O'Connell, Suzanne Salsman

We go to you - physically

ALS: A QUICK GUIDE FOR OCCUPATIONAL THERAPISTS (OTs)

Centre de réadaptation STAN CASSIDY Centre for Rehabilitation
Towards Independence / Vers l'autonomie

Head and neck support in wheelchair and bed

Vista Collar

Headmaster Collar

	PROS	CONS
Vista collar	More rigid for increased support and most popular. Pads are moisture-wicking and replaceable. Adjustable heights.	One size fits all (6 sizes in one), more obtrusive appearance.
Soft collar	If client cannot tolerate rigid collar.	Provides less support, less durable.
Headmaster collar	Breathable, firm support. Many sizes and colours available. Easy to clean, more subtle appearance.	Harder to adjust – minimal adjustments are possible by bending the collar. May need pad over collarbone.

Shoulder support: Prevent subluxation and pain

- To be worn when ambulating
- Try different options, some are more cumbersome
- Some clients find kinesiotaping helpful initially
- Other shoulder supports: https://www.neurorehabdirectory.com/product-category/shoulder-subluxation-sling
  - Rolyan Custom Hemi Arm Sling
  - Rolyan MFC Unilateral Shoulder Orthosis
  - Wilmer Orthosis
- In sitting:
  - Use a breastfeeding pillow to support arm/shoulder when:
  - Sitting in bed or recliner
  - Travelling in car
  - Using electronics and while reading

Photo courtesy of Ottobock

Hands

- Resting hand splint: to maintain ROM and prevent contractures
- Restorative hand splint has better compliance than custom-molded due to softness
- Wrist splint can help with grasp
- Thumb spica splint can be used when muscle wasting is apparent in thenar eminence to assist with grasp

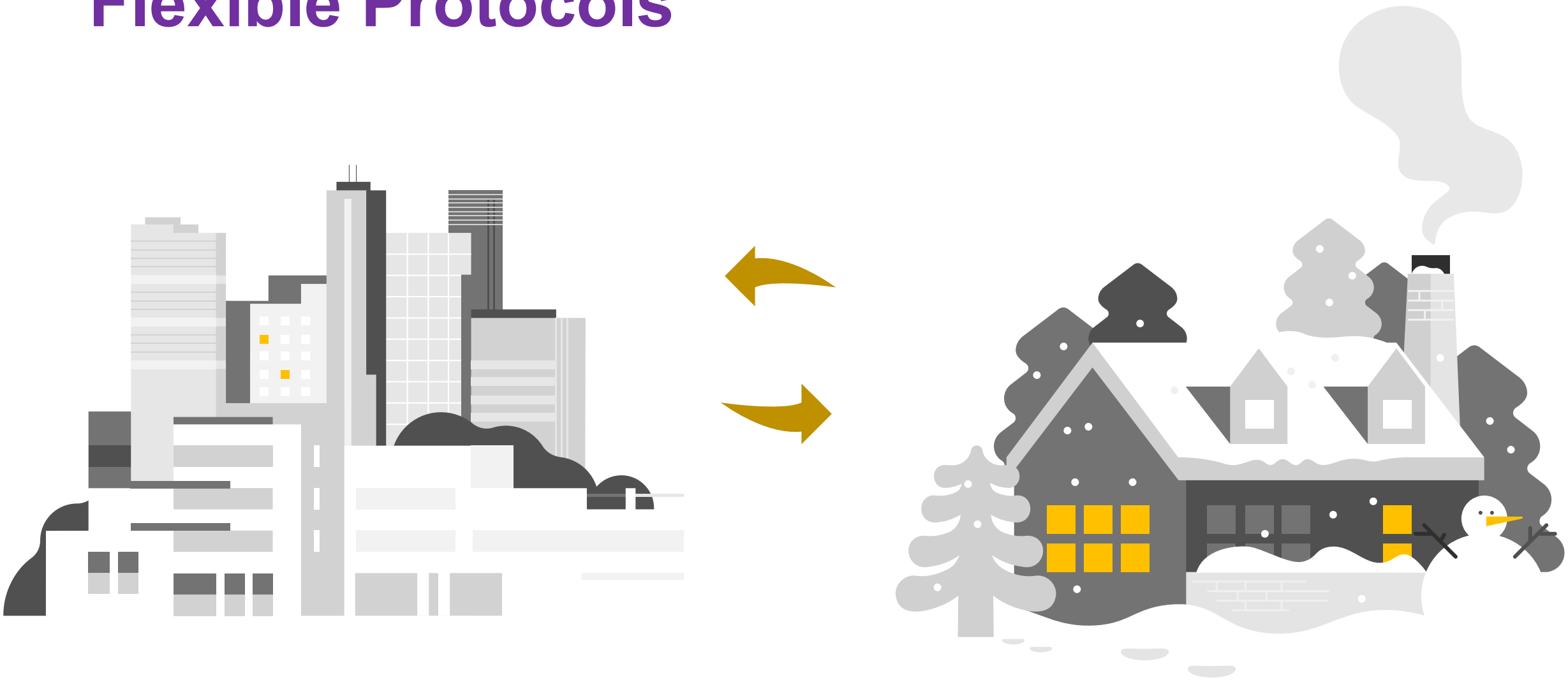
Feet

- Consult physiotherapist for appropriate recommendations for gait
- Dictus band raises foot after toe-off and reduces risk of tripping
- Night splint: to prevent foot drop at night, maintain ROM for wheelchair positioning, prevent pressure sores and contractures, and improve ambulation

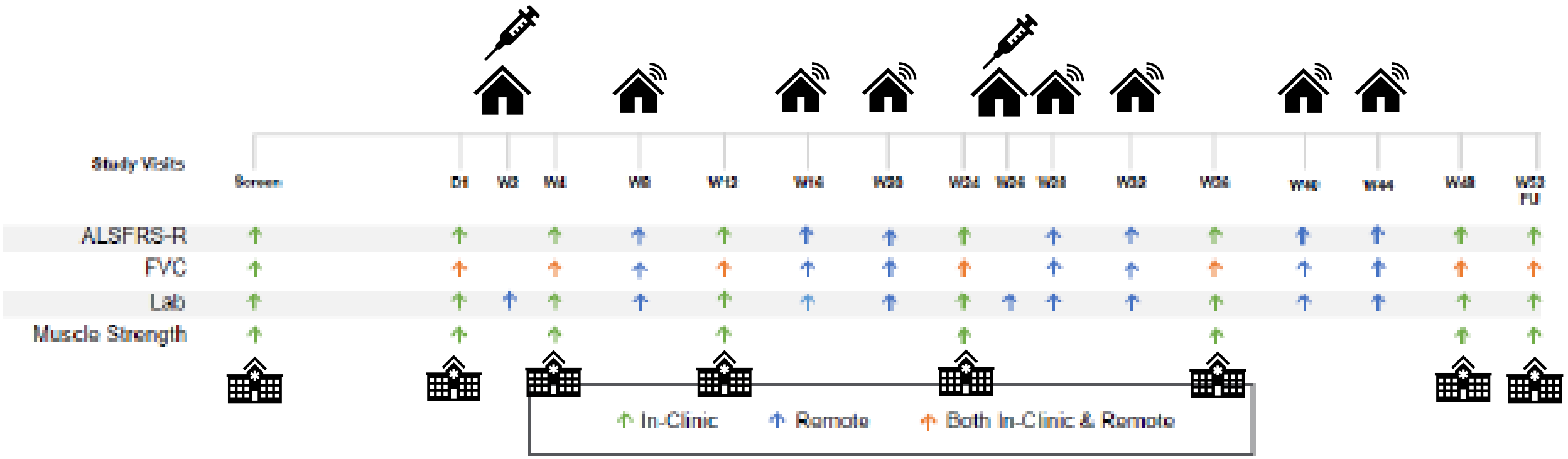
Dictus Band

Pictures are for illustrative purposes only

Flexible Protocols



Minimize Travel



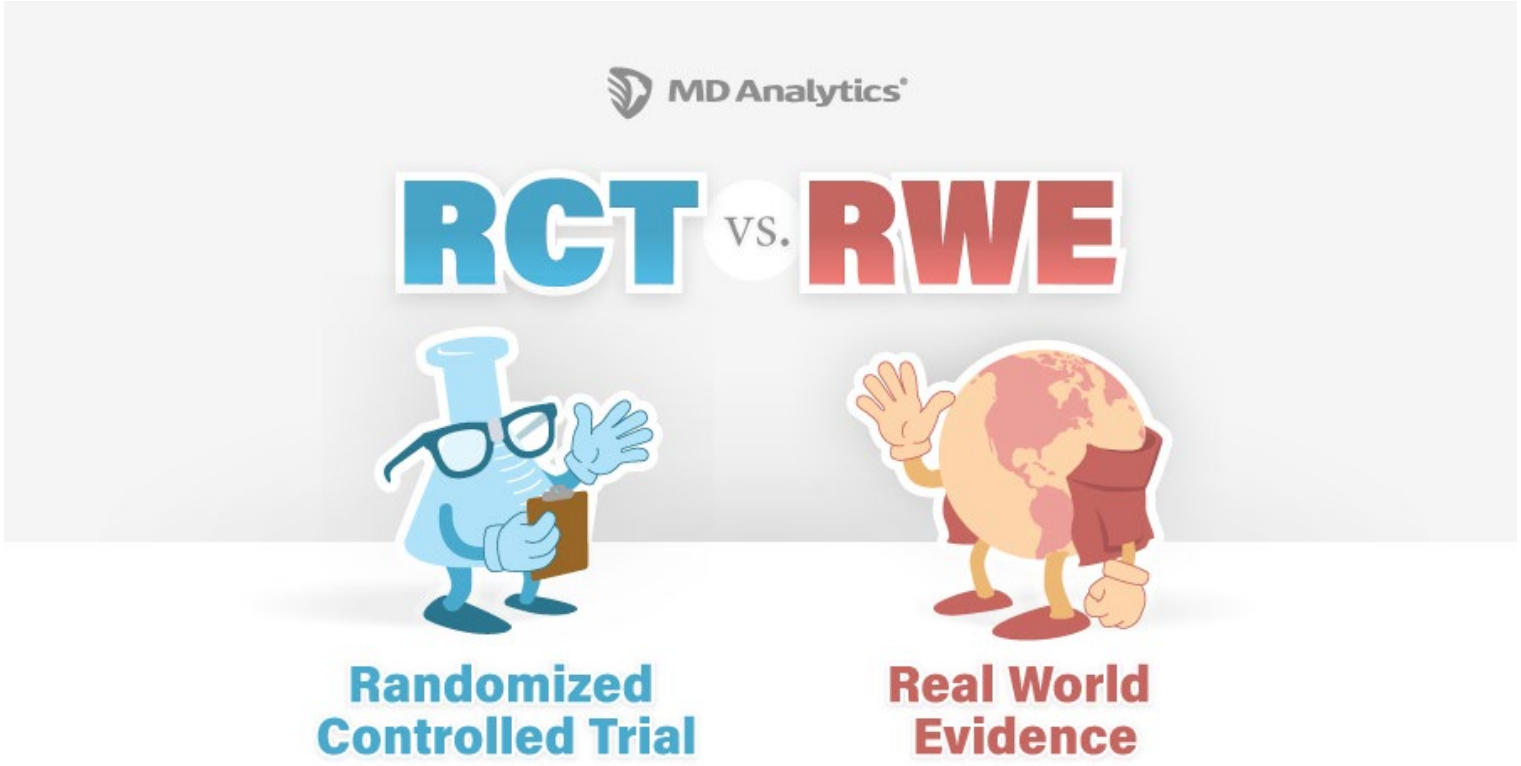
The Canadian Neurologic Disease Registry

A Multi-centre, National Collaborative Study

- Clinic-based recruitment & prospective data collection
- Over **5400 patients** registered across all neuromuscular disease
- Initiated 2010; continual growth & expansion
- Clinical data collection in DMD, SMA, DM, LGMD, ALS, CMS

38 NM clinics
14 ALS clinics

136 investigators



Registries and Real World Evidence

Patient Perspectives

“All people with this illness have value and their lives depend on acquiring the necessary resources to make sure they are able to use their value”

How to Break the News in ALS/MND

A Primer for Physicians and Allied Health Professionals

ALS, amyotrophic lateral sclerosis, MND: motor neuron disease

A-L S-PIKES Protocol for Delivering News in ALS/MND (at diagnosis and throughout the disease course)

A Advance Preparation

L S Location & Setting

P Patient's Perceptions

I Invitation

K Knowledge

E Empathy/Emotions

S Strategy & Summary

This program will focus on delivering news in the early stage of ALS/MND (i.e., delivery of diagnosis and "aftermath" period). However, these communication skills can be used at other time points throughout the disease trajectory by all ALS/MND team members.

What needs to be communicated at different stages differs, but the *skills for communicating* are essentially the same.

This protocol has been adapted from the SPIKES protocol developed for delivering oncologic diagnoses.

ALS, amyotrophic lateral sclerosis; MND, motor neuron disease

Adapted from: Baile W, et al. Oncologist 2000;5:302-311. Buckman R. Community Oncology 2005;2:138-42. EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis. Eur J Neurol. 2012;19:360-75.

Summary

ALS is a progressive, heterogeneous disease which complicates prediction of long-term outcomes[1]

Integrate palliative care into routine management before end of life stages

Multidisciplinary care for patients with ALS optimizes healthcare delivery and prolongs survival[9]

Disease-modifying therapies and strategies for symptom management are important for the overall care and quality of life of patients with ALS

1. Canadian Organization for Rare Disorders' (CORD) response to the PMPRB consultation 2018; 2. Statland JM et al. *Neurol Clin*. 2015;33(4):735–48; 3. Joyce NC, Carter GT. *PM R*. 2013;5(5 0):S89–95;
4. Bock M et al. *Neurol Clin Pract*. 2017;7(6):488–98; 5. Dziewas R et al. *Dysphagia*. 2017;32(1):78–82; 6. Motor neurone disease: assessment and management. National Institute for Health and Care Excellence. 2016.
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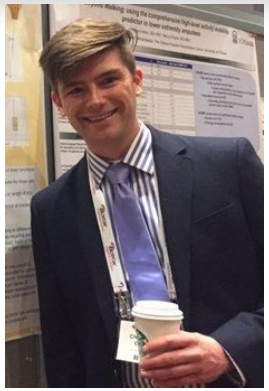
https://als.ca/wp-content/uploads/2017/02/A-Guide-to-ALS-Patient-Care-For-Primary-Care-Physicians-English.pdf. Accessed 22 September 2020; 8. Finegan E et al. *Front Neurol*. 2019;10:260; 9. Hardiman O et al. *Nat Rev Neurol.* 2011;7(11):639–
49; 9. Miller RG et al. Neurology. 2009;73(15):1227–33.

ALS Clinic Team









Thank you!

Resources:
https://als.ca

https://www.als-mnd.org/support-for-health-professionals/how-to-break-the-news-in-als-mnd