

# Living a quality life with a neurodegenerative diagnosis

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# Objectives

- 1 – Explain the importance of interprofessional collaboration in supporting a quality of life in persons with neurodegenerative disorders (CANMED role: Collaborator)
- 2 – Identify strategies to optimize early engagement of interprofessional palliative care in neurodegenerative disorders (CANMED roles: Health Advocate, Leader)
- 3 – Describe interventions to manage physical, social and emotional needs of persons and families living with neurodegenerative diagnoses (CANMED role: Medical Expert)

# Caveat

**Health-Related Quality of Life**

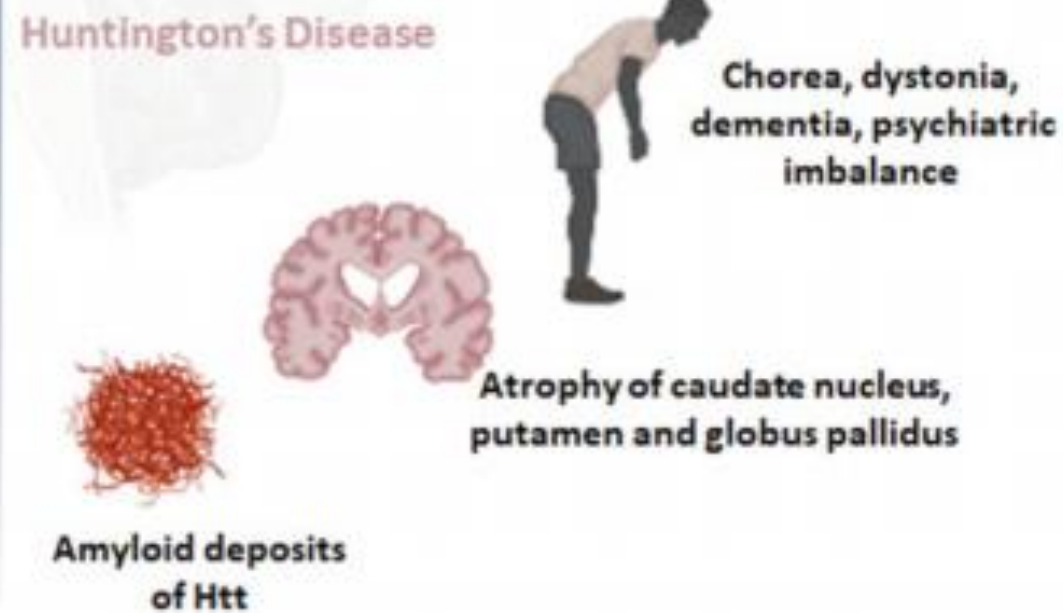
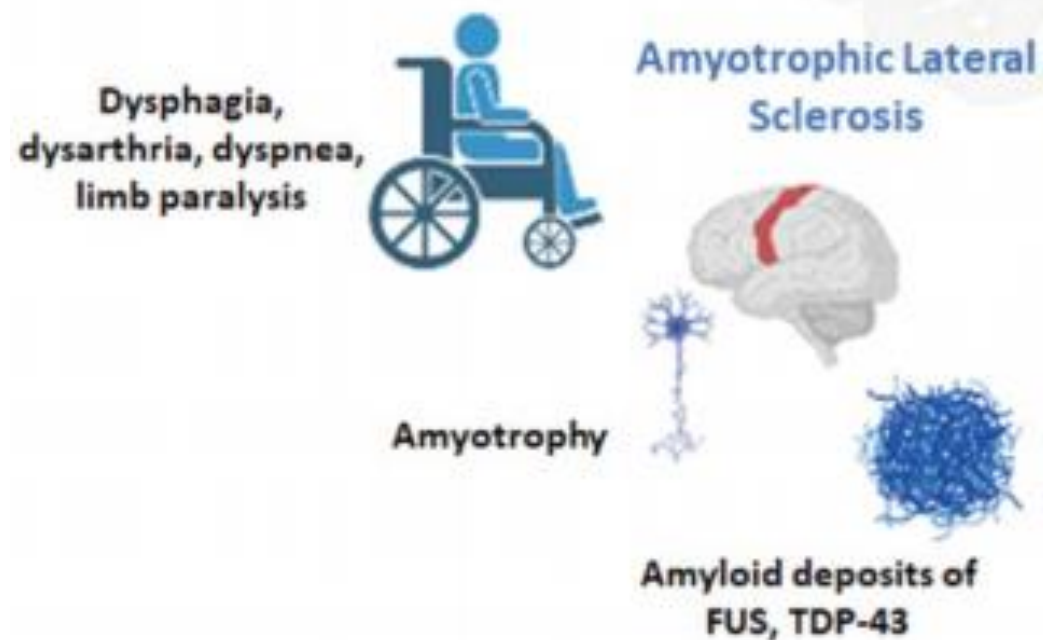
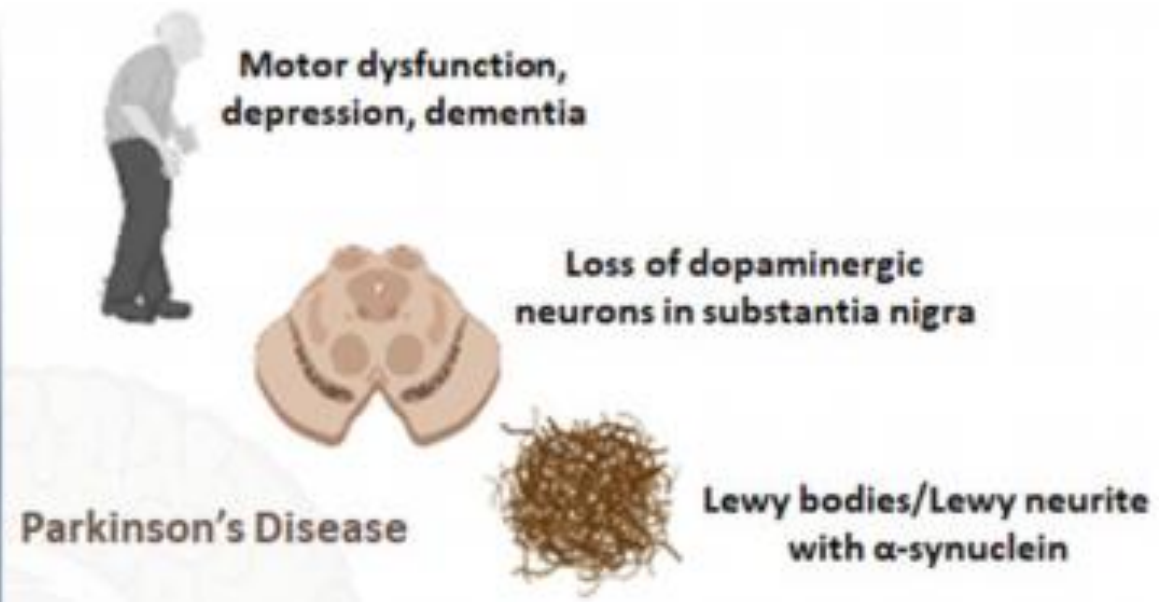
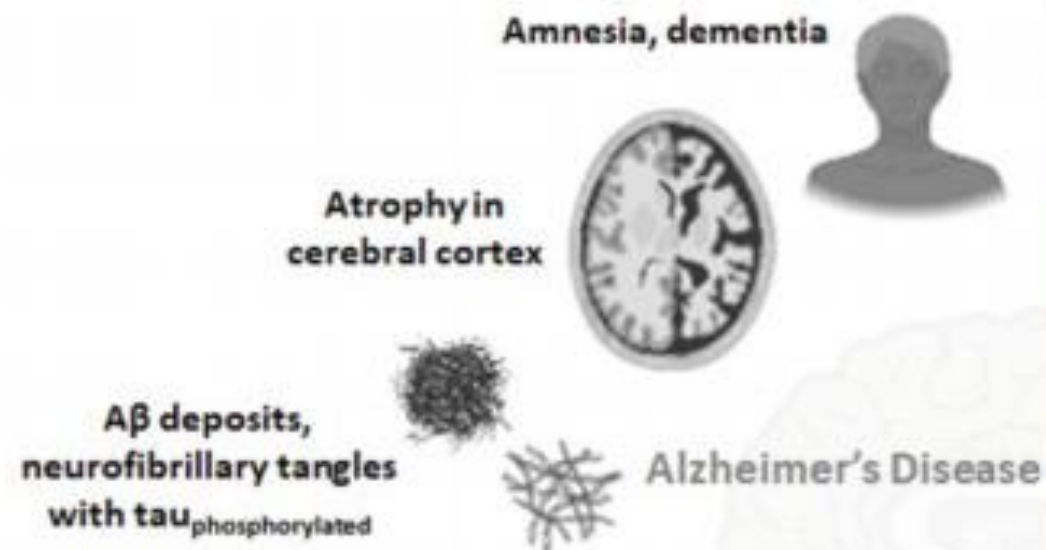


**Quality of Life**



# Neurodegenerative Conditions

General overview of more common neurodegenerative disorders, incidence, symptoms and progression



# ALS: a progressive, heterogeneous disease

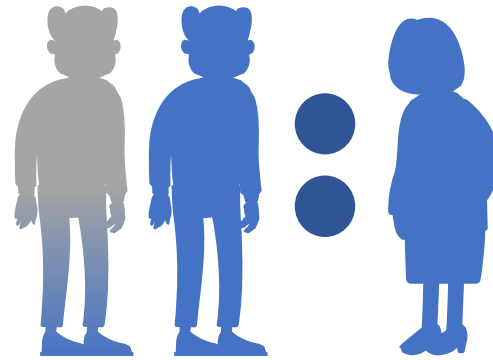


**3,000**  
live with  
ALS<sup>1,2,4</sup>

**1,000**  
die from ALS  
each year<sup>4</sup>

**2/100,000**  
new cases of  
ALS each year<sup>2</sup>

Mean age of onset 55–65 years<sup>2</sup>



**1.5 male : 1 female<sup>2</sup>**

**80%**

Die within 2–5 years  
of diagnosis<sup>3</sup>

**10%**

Survive >10 years<sup>3</sup>

**1:350**

Lifetime risk among men<sup>5</sup>

**1:500**

Lifetime risk among women<sup>5</sup>

Respiratory complications is the most common cause of death in patients with ALS<sup>6</sup>

**While there is no cure, there are treatment options available to improve quality of life, function and survival<sup>1</sup>**

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](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](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.

# What happens to people with ALS

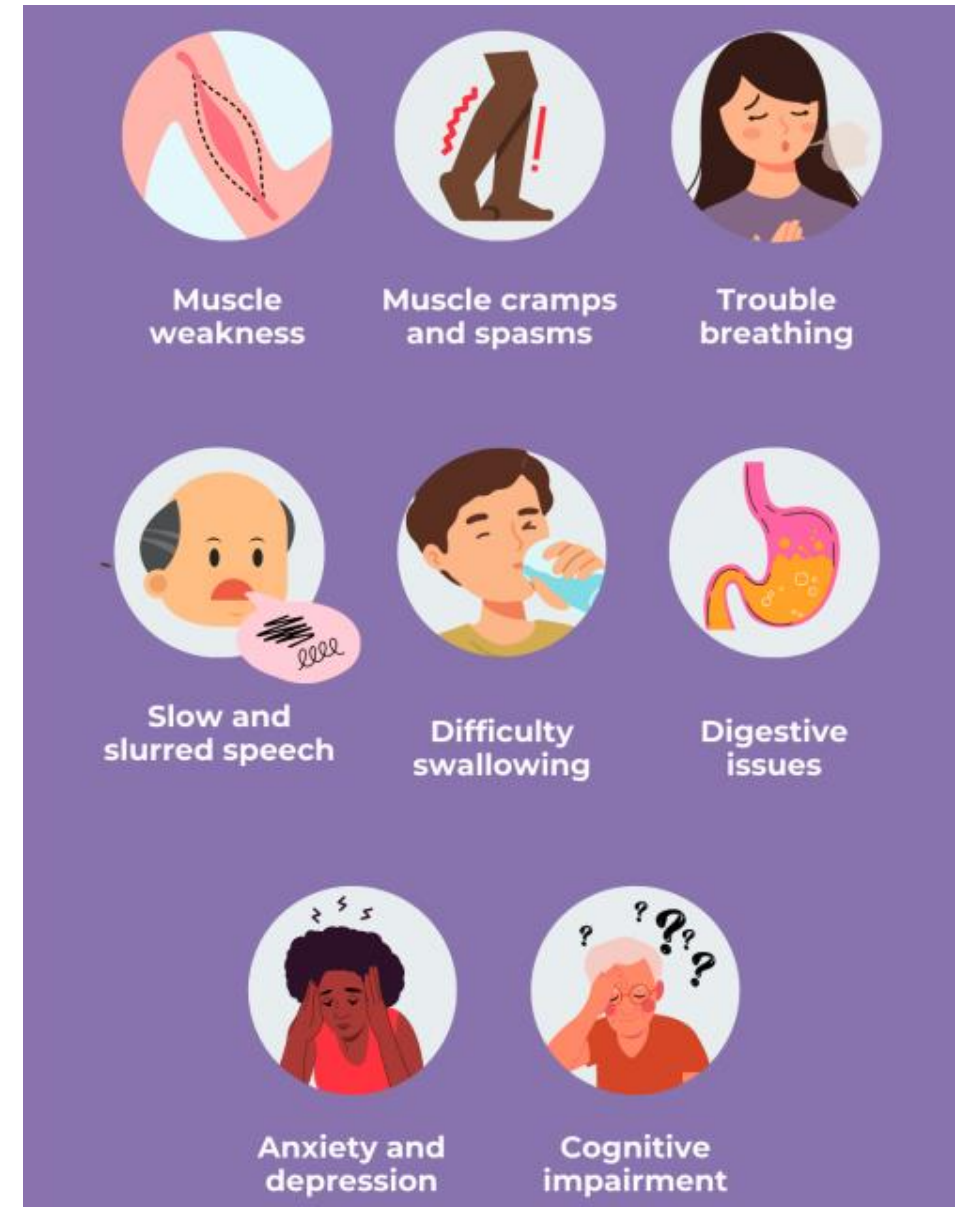
## •Upper motor neuron:

- Weakness
- Spasticity, hyper-reflexia
- Emotional lability
- Loss dexterity
- Slowed movement



## •Lower motor neuron:

- Atrophy
- Weakness
- Hypo-reflexia
- Muscle cramps
- Fasciculations



# Parkinsons



- 17.4 per 100,000 between the ages of 50-59 and 93.1/100,000 between the ages of 70-79
- middle-aged and elderly
- 1.5x in men
- >100,000 in Canada prevalence



# Parkinsons Disease – Late Symptoms

- Sleep disorder – 100%
- Depression 40–60%
- Dementia – 20-75%
- Pain – up to 50% (under reported)
- Hallucinations/psychosis – 25-30%
- Severe dysphagia
- Orthostatic Hypotension
- Dysarthria, hypophonia, tachylalia, and freezing of speech



# Parkinsons Progression

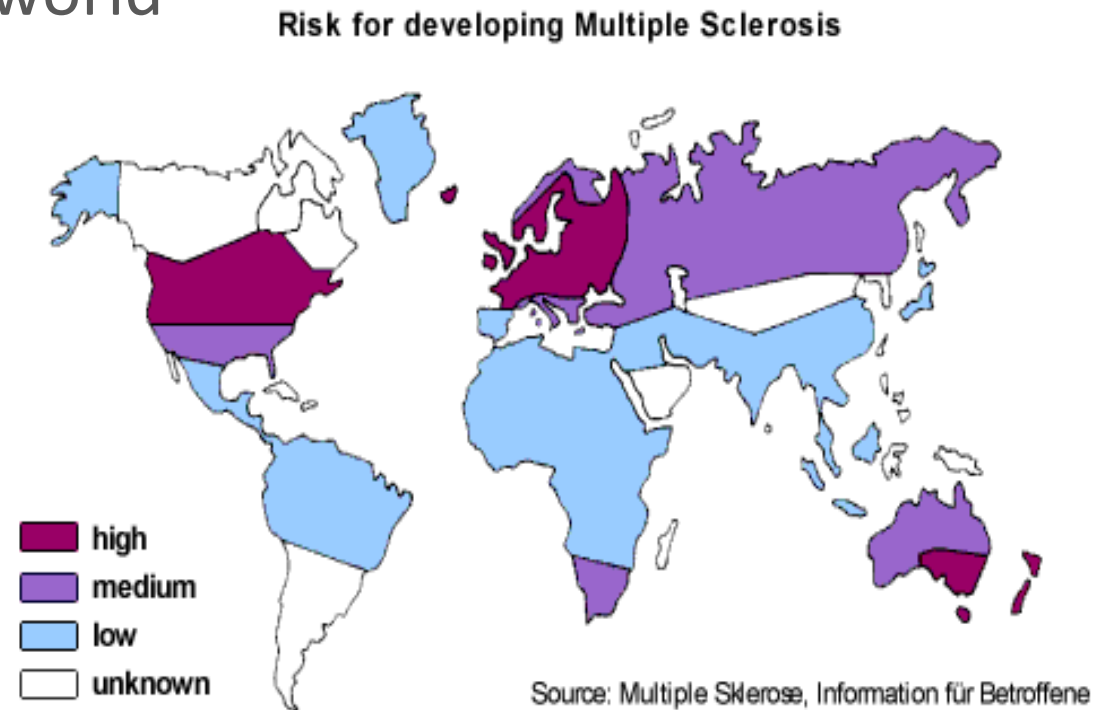
- mean duration of disease was 14.6 years.
  - diagnosis 1.5 years
  - maintenance 6 years
  - complex 5 years
  - palliative care 2.2 years.
- Mean time disease onset to wheelchair-dependence est. 14 years
- about a third have relatively milder disease and remain stable for many years



Andy Seward had to be helped by security officials as he carried the torch in North Devon (Picture: BBC)

# Multiple Sclerosis

- Most common neurologic disease of young adults
- 3 times more common in women
- Canada has one of highest rates in the world
- 1.8-3.5/1000
- Over 90,000 in Canada



Source: Multiple Sklerose, Information für Betroffene  
by Günter Krämer and Roland Besser  
Georg Thieme Verlag Stuttgart

# Most Common Symptoms

- Sensory – 82% 5 yrs 100% life
- Spasticity – 75%
- Weakness – 100% lifetime
- Fatigue – 75%
- Pain – 30%
- Depression- 30%



Photo from <http://abcnews.go.com/Health/top-10-myths-multiple-sclerosis/story?id=19752065>

# Natural history of multiple sclerosis

25 year study: 80% of patients developed secondary progressive disease, 15% died, 65% required walking aid

- Walking impairment at 8yrs,
- Cane at 20yrs
- Wheelchair at 30yrs

Confavreux C, Vukusic S. 2006

# What's Unique...

- Progressive deterioration in function
- Communication and cognition involvement
- Series of losses and grief
- No known cures



# Neuropalliative and rehabilitation care



*“a holistic approach to the care of neurological patients with significant disability, complex needs, and a potentially shortened life-span. It is patient-centered and involves diagnosis of clinical problems at all stages, rehabilitation to maintain function, care coordination and appropriate palliation to relieve symptoms”*

# The Science

Literature and evidence in palliative care and quality [of] life for persons living with neurodegenerative conditions



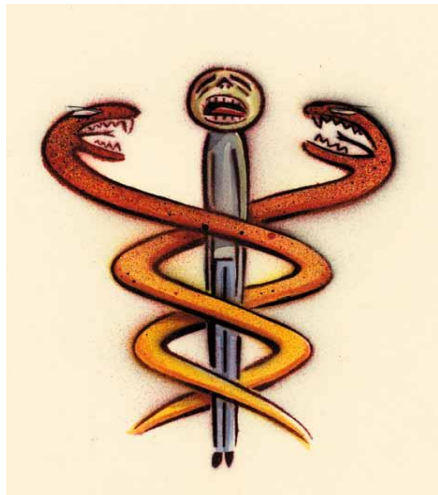
# The Needs

Mobility

Swallowing

Speech

Pain



Financial

Isolation

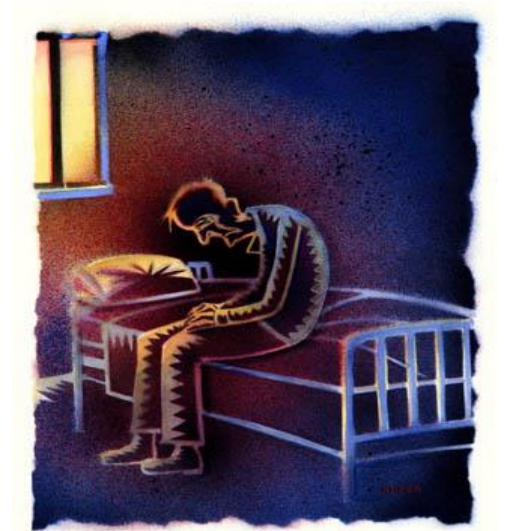
Social



Meaning of life

Hope

Spiritual



Veronese, S., Gallo, G., Valle, A., Cugno, C., Chiò, A., Calvo, A., ... Oliver, D. J. (2015). The palliative care needs of people severely affected by neurodegenerative disorders: A qualitative study. *Progress in Palliative Care*, 23(6), 331-342. <https://doi.org/10.1179/1743291X15Y.0000000007>

# Palliative Care improves Quality of Life

Meta-analysis of RCTs: Palliative care associated with improvements in quality of life

RCT: Patients with progressive neurological disease who received specialized palliative care: outcomes better for symptom burden, emotional and social function, spiritual, QoL

Bužgová, R., Kozáková, R. & Bar, M. The effect of neuropalliative care on quality of life and satisfaction with quality of care in patients with progressive neurological disease and their family caregivers: an interventional control study. *BMC Palliat Care* **19**, 143 (2020). <https://doi.org/10.1186/s12904-020-00651-9>

Kavalieratos D, Corbelli J, Zhang D, Dionne-Odom JN, Ernecoff NC, Hanmer J, Hoydich ZP, Ikejiani DZ, Klein-Fedyshin M, Zimmermann C, Morton SC, Arnold RM, Heller L, Schenker Y. Association Between Palliative Care and Patient and Caregiver Outcomes: A Systematic Review and Meta-analysis. *JAMA*. 2016 Nov 22;316(20):2104-2114. doi: 10.1001/jama.2016.16840. PMID: 27893131; PMCID: PMC5226373.

# Specialized Palliative Care for complex cases

Severe patients fast tracked to specialized palliative care teams had improved QoL, pain, respiratory and sleep outcomes

Individualised care that supports engagement in decision-making and therefore spiritual well-being is essential for promoting patient empowerment, autonomy and dignity.

Veronese S, Gallo G, Valle A, *et al* Specialist palliative care improves the quality of life in advanced neurodegenerative disorders: NE-PAL, a pilot randomised controlled study *BMJ Supportive & Palliative Care* 2017;**7**:164-172.

Paal P, Lex KM, Brandstötter C, Weck C, Lorenzl S. Spiritual care as an integrated approach to palliative care for patients with neurodegenerative diseases and their caregivers: a literature review. *Ann Palliat Med*. 2020 Jul;**9**(4):2303-2313. doi: 10.21037/apm.2020.03.37. Epub 2020 Apr 7. PMID: 32279521.

# The Art

Practical ideas, tips and strategies to support quality [of] life for those living with neurodegenerative conditions

# Practice Parameters - Guiding Principles



High priority on self-determination



Timely information for decision making



Decisions are dynamic and can change



Team should address full continuum of care

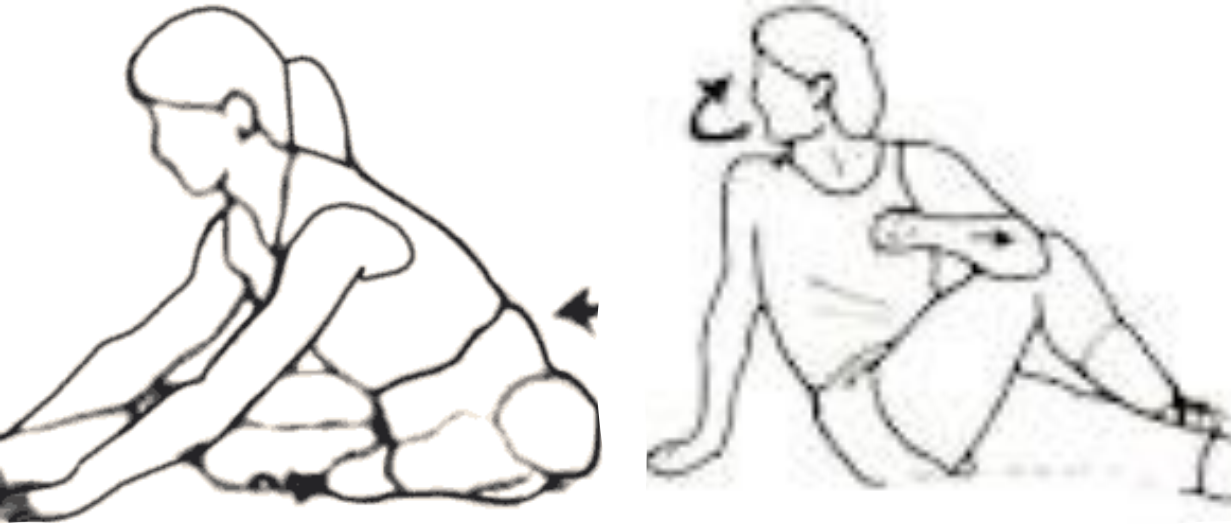


# Pain

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- Immobility
- Contractures
- Skin
- Positioning - camptocormia
- Central neuropathic
- Peripheral neuropathic
- Infections
- Spasticity/spasms/Rigidity
- Dykinesias/dystonia

*If I woke up  
in the  
morning  
and nothing  
hurt,  
I would think  
I was dead.*



# Pain Treatment

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- Stretching, exercise program
- Positioning
- Meditation
- TENS, massage, modalities
- Desensitization techniques





# Mobility and Everyday Life

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## ALS: A QUICK GUIDE FOR OCCUPATIONAL THERAPISTS (OTs)

### Head and neck support in wheelchair and bed



Vista Collar



Headmaster Collar

	PROS	CONS
<b>Vista collar</b>	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.
<b>Soft collar</b>	If client cannot tolerate rigid collar.	Provides less support, less durable.
<b>Headmaster collar</b>	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

# Energy Management Strategies



Prioritize



Plan



Pace



Position

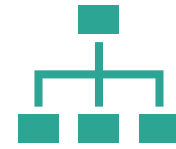
# Energy Management Strategies



Do



Delay



Delegate



Dump

# Exercise

- Moderate-intensity is probably beneficial for function and QoL (B)
- Personalize strength (submax effort) and aerobic training common-sense approach
- Regular stretching/ROM – evidence for spasticity (C)

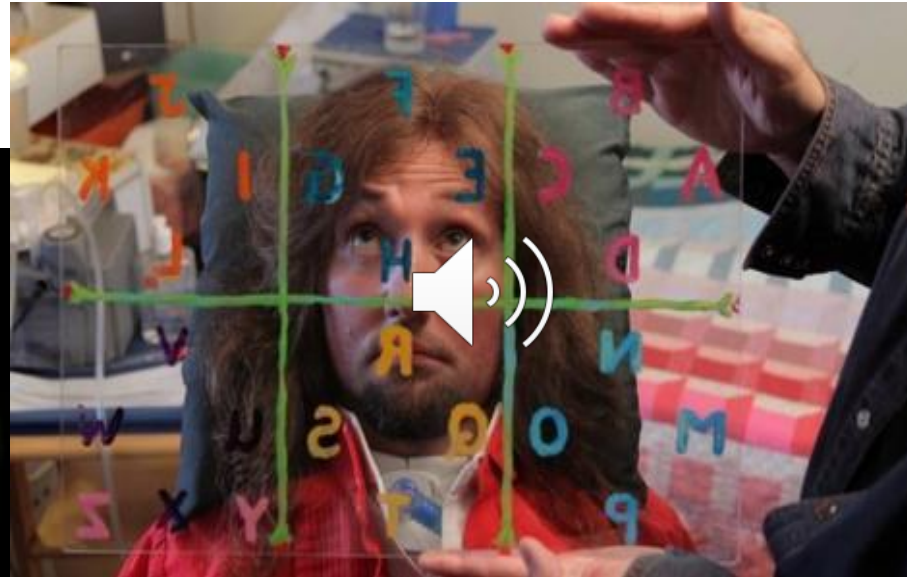


# Communication

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- *Not being able to speak, does not mean having nothing to say*
  - Teach non-verbal strategies early
  - Digitally record voice
  - Evaluate skills, resources, motor and cognitive abilities; referral to OT, Aug Comm, Environmental Control
  - High tech and low tech
  - Scanning techniques, family and caregiver education, signals
  - There are no studies to support any particular treatment

# Augmentative and Alternative Communications



# Bronchial Secretions and Saliva

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- Stimulation of b-adrenergic receptors produces thick protein and mucus-rich secretions :  
guaifenesin, beta-blocker, N-acetylcysteine
- Stimulation of cholinergic receptors produces thin serous secretions : anticholinergic  
bronchodilator
- Saliva – anticholinergic first line
- Suction Machine, Airway clearance techniques (Cough Assist)
- Oral Care

# How do patients feel?

**Fear of  
'choking to  
death**

*"I woke up and felt like I couldn't breathe, I tried to cough but couldn't. I got so scared and panicked..."*

"All night every hour we were up trying to get the phlegm up, we were both exhausted and frustrated"



# Lung Volume Recruitment – Airway Clearance

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# Mechanical Insufflator-Exsufflator

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# Social and Emotional

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- Determine goals, obstacles and facilitators
- Referrals as appropriate: psychology, spiritual counselor
- Identify medical needs (depression, pseudobulbar, cognitive)
- Community partners
- Autonomy and respect
- Family needs

## Clothing

See the following websites for adaptive clothing that will serve you better!

### Silverts:

Easily shop by specific condition: or by mobility level. Clothing is separated into A+ and S+, A+ being for people who require assistance dressing, and S+ for people who dress themselves. They also have a blog on their website with articles including how to shop for others, health tips, and more!  
Examples include: full side zip pants, open-back gowns and shirts, wheelchair pants, anti-strip jumpsuits  
Sizing: small-2XL  
[www.silverts.com](http://www.silverts.com)



### Easy Wear Adaptive Clothing:

This is an independent retailer based in Ontario. Sells wheelchair adaptive clothing, shirts, pants, dresses and women's undergarments.  
Sizing: small-4XL  
[www.easywearadaptiveclothing.com](http://www.easywearadaptiveclothing.com)



Men's Long Sleeves Polo's & Flannels - Medium 40-42

## Clothing

### Healthwick:

Specializes in clothing as well as surgery and injury recovery products. Includes mobility aids, incontinence/catheters, liquid nutrition/thickener and wound cleaning. They also sell slippers and anti-slip socks. Their website features informative pages explaining how to use their products with videos.  
Sizing range: XS-3XL  
Note: Fewer colour and style options on this website than others.  
[www.healthwick.ca](http://www.healthwick.ca)



### Gerri Fashions:

Sells pants, tank tops, shorts, and more. Made in Canada and shipped from Ontario. Also sells capes, canes, slippers, socks and underpads. This company sells more formal clothing than the others.  
Size: XS-3XL  
[www.gerifashions.com](http://www.gerifashions.com)



## Hair Products

Styling and washing your hair doesn't have to be complicated. These products and many more can simplify your routine!

This 3-barrel curling iron called "Trademark Beauty's Babe Waves" can be used with one hand. Just clamp onto the hair you would like curled! It can be purchased on Amazon.



Carol's Daughter Shampoo makes a water-to-foam shampoo that can be applied directly to the scalp, requiring only one hand. Because of its water-based formula it is easy to dispense! It can be purchased on Amazon, at Shoppers Drug Mart, or Walmart.



### Kohl Kreatives.com

This UK company has created a makeup brush collection specifically for those with motor disabilities. All brushes stand by themselves and are easy to grip. In addition to their brushes, they also sell "quickie stickies", stickers that you can place on your face to aid with contouring, eyeshadow, eyeliner and more.



## Makeup

### Rare Beauty:

Rare Beauty products have features that simplify opening and application, with mobility issues in mind. For example, the spheres on top of the applicators of their base products and liquid highlighters make it easier to open and hold when applying. They also have a very inclusive colour range. This brand is relatively new but is gaining popularity quickly, so more products are likely to be available soon!

[Rarebeauty.ca](http://Rarebeauty.ca)



### Guide Beauty

This company created the "Guide Wand", an eyeliner applicator that is specifically designed to be easier to grip and use. They also sell eyeshadow brushes, mascara, and eyebrow gel with a similar design. They currently only ship to the United States, however, they are looking to expand to Canada soon. Check their website periodically if you're interested!

[www.guidebeauty.com](http://www.guidebeauty.com)



Guide Eyeliner Duo

## Footwear

### Billyfootwear.com

This shoe company creates shoes with wrap-around zipper closures to make it easier to put on shoes. They also make it easier to put a brace on if required. A pair also comes with removable insoles for adjustable depth. They also have a wide variety of colours and styles!



### Nike Flyease, nike.com

This shoe was designed specifically with mobility issues in mind. The entire heel (including the sole) of the Nike Go FlyEase pivots open for a totally hands-free entry.



More Questions? Talk to your occupational therapist!



# TRAVELLING WITH A WHEELCHAIR:

Things to consider...



## DO YOUR RESEARCH

Things to consider before booking a trip:

- Experience (relaxation, exploration, sight seeing, etc...)
- Location
- Travelling (flights, living arrangements, transportation, etc...)
- Accommodations

Here is a great website to help you discover accessible destinations:  
<https://accessnow.com/>

## PACKING

Ensure to pack all the necessities in your carry on bag.

These may include:

- catheters
- medication
- change of clothes
- small set of tools and chargers
- Transfer board
- Wheelchair instruction manual
- Bring pillows or whatever you need to be comfortable and safe

In your suitcase/checked bag:

- spare cushion (if you have one)
- **\*\*Consider baggage weight limit\*\***

## ARRIVAL AT AIRPORT

- Arrive at least **2 hours** in before your flight
- (**3 hours** is recommended for international flights)
- At check-in, present yourself and remind clerk of your accessibility needs.
- A crew member can help you go through security and gate.
- You will not be required to get out of your wheelchair during security checks.
- Officers will have to pat you down or use a metal detector when going through security.

## BOOKING A FLIGHT

- Request a wheelchair service. The request can be made when booking your flight online. You may also contact the airline directly for this request.  
**\*Notify airline of any special assistance request\***

- Give airline at least **48h notice** of your accessibility needs.

**\*\*It is recommended to call your airline immediately after booking to confirm your accessibility needs\*\***



- Provide info about your wheelchair including the dimensions, weight, and if it is a manual or power chair.
- If you have a connecting flight, ensure you have a long layover (**at least 1 hour and a half**).

## AIRLINES CAN HELP WITH THE FOLLOWING:

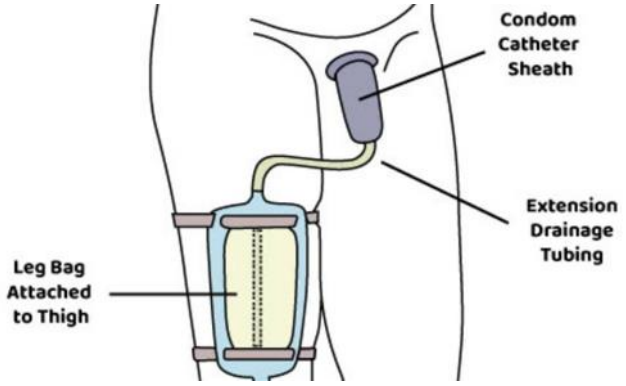
- Checking in at check-in counter
- Proceeding through security screening
- Proceeding to boarding area after check-in
- Boarding and disembarking
- Storing and retrieving carry-on baggage
- Accessing the largest washroom available on aircraft
- Proceeding through border clearance
- Retrieving checked baggage
- Proceeding to general public area upon disembarkation, to a location where terminal personnel may assist you with proceeding to curbside zone or finding your connecting flight
- Free extra carry-on bags
- If available, you can request a discounted fare for your caregiver
- And more!

**\*Please note that all airlines have different policies concerning services for passengers with disabilities.**





Virtual Reality



# Bring on the team

Should be initiated early in disease course

Quality of life/Symptom management

Discussions include:

- End of life decisions and directives
- Options for respiratory and nutritional support
- Inform on legal situation for advanced directives
- Formulation of advanced directives and plan

Collaborate with health team and family

Assist with grief and loss support



# Towards shared care

Opportunities for interprofessional and multidisciplinary care across the life course

# “Holistic Rehabilitative Palliative Care”

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‘All the work of the professional team .. is to enable the dying person to live until he dies, at his own maximal potential performing to the limit of his physical and mental capacity with control and independence whenever possible”.

- Cicely Saunders



# Principles of Rehab Palliative Care in PND

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Prospective person-centred care

Focus on Function beyond symptoms

Opportunities for Enablement and Self Management

Coordinated Continued Care

- Maximize Quality of Life

- Minimize Stress and Burden

- Maintain Function



- With compassion
- In partnership
- Through connection
- In a risk confident way

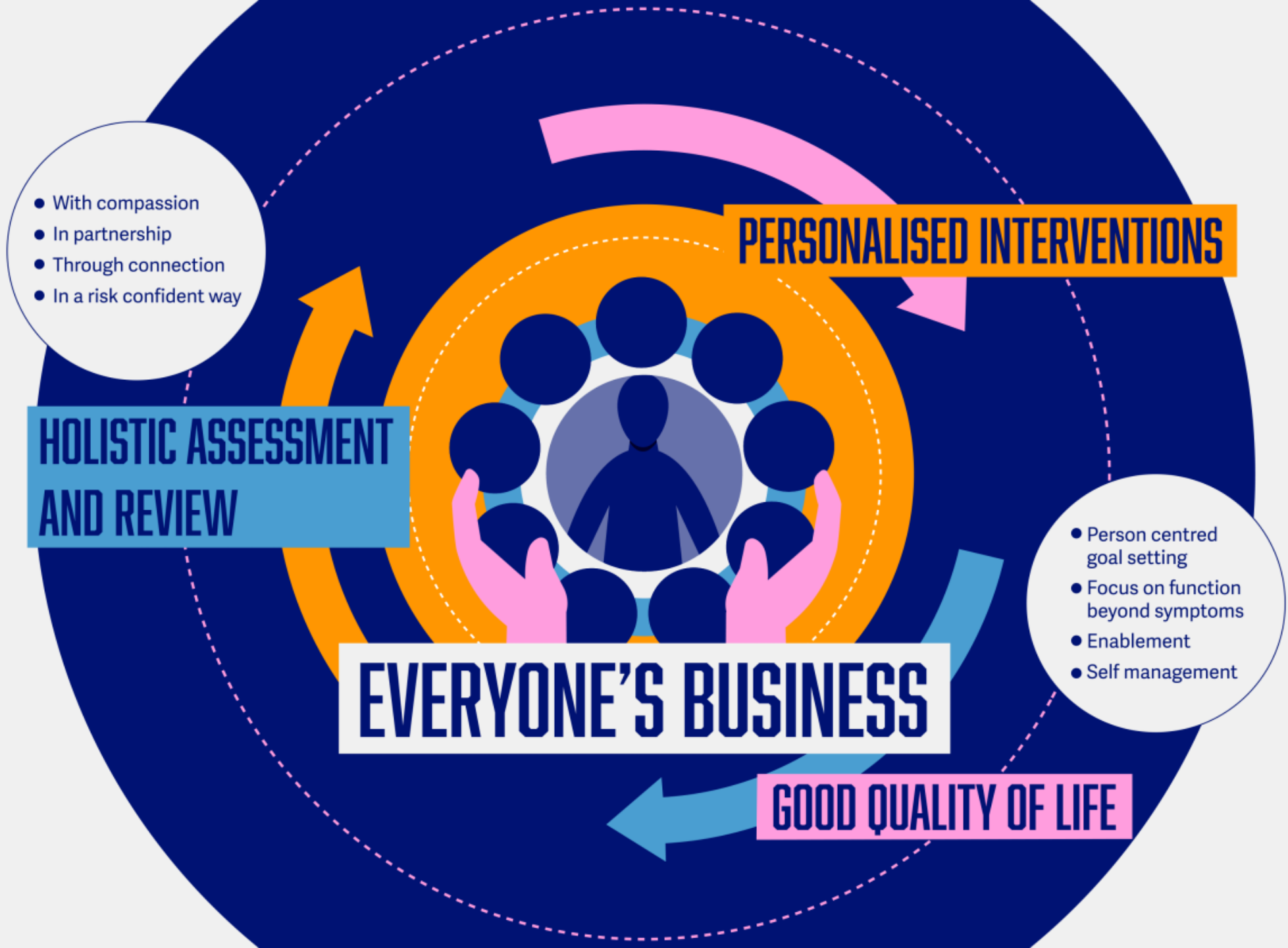
**HOLISTIC ASSESSMENT  
AND REVIEW**

**PERSONALISED INTERVENTIONS**

- Person centred goal setting
- Focus on function beyond symptoms
- Enablement
- Self management

**EVERYONE'S BUSINESS**

**GOOD QUALITY OF LIFE**



# Summary in Neurodegenerative Care

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**Palliative care** occurs from diagnosis to death

Quality of life objectives vary patient to patient

Focus on Function with interventions to maintain quality of life

Patient goals change from maximizing function to dignified death

Proactive care can reduce morbidity, maintain function, ease stress, and aid planning



*“...when this all started, I asked myself. ‘Am I going to withdraw from the world, like most people do, or am I going to live?’ I decided I’m going to live – or at least try to live – the way I want, with dignity, with courage, with humor, with composure”-*

*Morrie Schwartz* from Tuesdays with Morrie

- <https://filmfreeway.com/DancingWithALS>

