The Role of Physiotherapy in Treating ALS

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rehabilitare: Latin: “to provide again with means”

- Rehabilitation has been described as the aim to ”restore, compensate, prevent or slow deterioration in functioning (sensory, physical, intellectual, mental, cognitive, or social) to help individuals to reach their optimal levels.”

Duttine A, Battello J, Beaujolais A, Hailemariam M, Mac-Seing M, Mukangwije P, et al. (2016) Introduction to Rehabilitation Factsheet. Handicap International. Available from: https://humanity-inclusion.org.uk/sn_uploads/document/2017-02-factsheet-rehabilitation-introduction-web_1.pdf

- The physiotherapist as an integral part of the multidisciplinary team is to rehabilitate, to optimise a healthy, functional, quality life. Our skill set enables us to assist the person **to feel at greater ease in a body that is rapidly changing.**

ALS

- Each individual with MND progresses at a unique rate.
- The different clinical phenotypes, the variable presentation, the variability in prognosis and the changing nature of the illness over time will impact these tailored requirements.
- Understanding the stages of the illness can serve as a wider framework to create intervention strategies and develop more comprehensive treatment plans

Dal Bello-Haas (1998) proposed stages that focus less on symptom presentation and more on functional independence and intervention consideration.

Stage 1

- Preserved functional independence,
- Muscle weakness with changes in resistance.
- Physiological support and continuity of normal physical activities are recommended.

Stage 2

- Involvement of the distal musculature
- Use of orthoses
- Maintenance
- Cardiopulmonary and neuromuscular conditioning and physical-functional training

Dal Bello-Haas (1998) proposed stages that focus less on symptom presentation and more on functional independence and intervention consideration.

Stage 3

- Moderate functional limitations
- Susceptibility to fatigue.
- Wheelchairs
- Orthoses for the arms.

Stage 4

- Severe phase
- Changes in the lower limbs.
- Continuation of the exercises
- Exercises aimed at preventing muscle contracture
- Positioning

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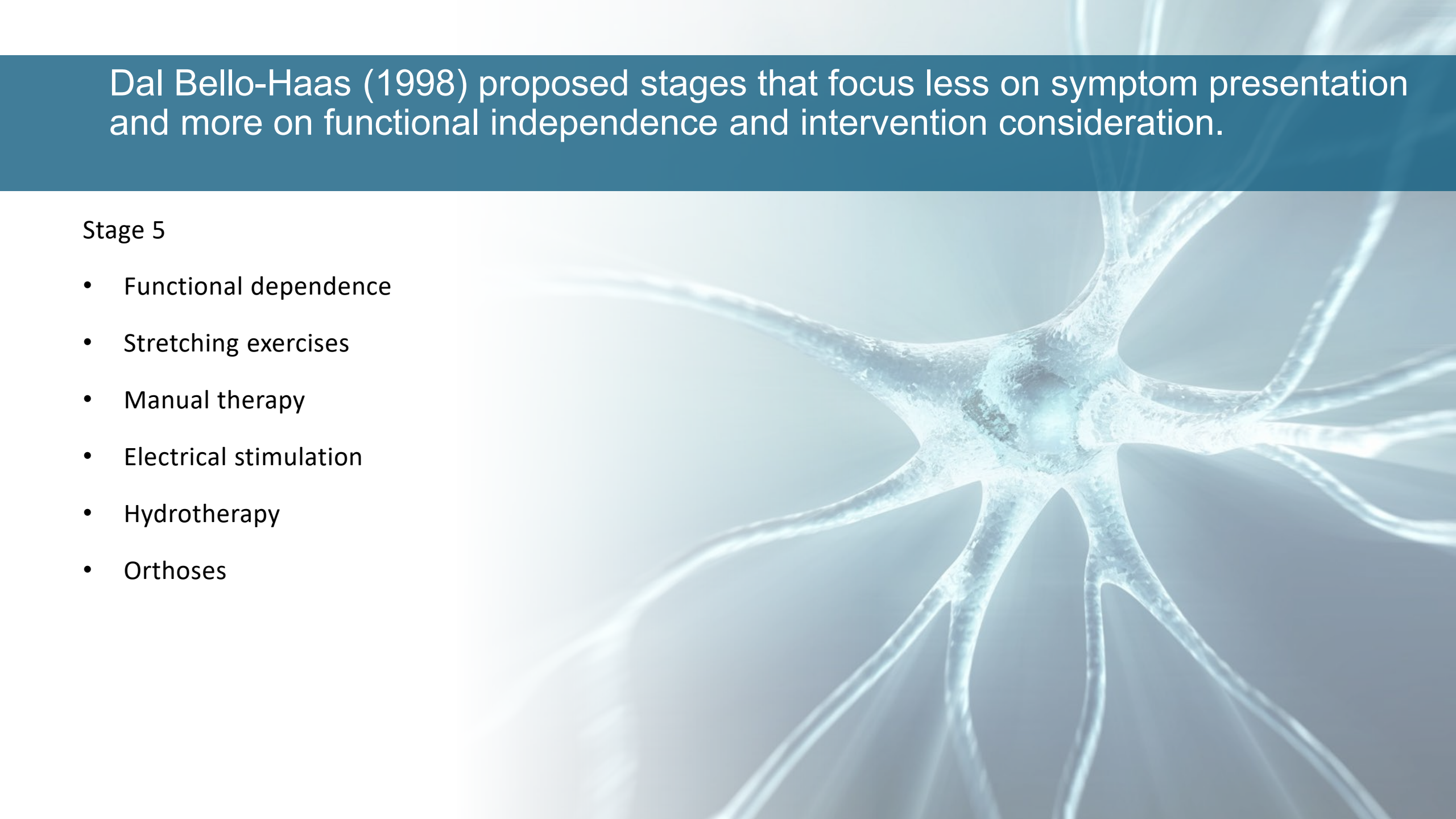


Stage 5

- Functional dependence
- Stretching exercises
- Manual therapy
- Electrical stimulation
- Hydrotherapy
- Orthoses

Dal Bello-Haas (1998) proposed stages that focus less on symptom presentation and more on functional independence and intervention consideration.

Stage 6

- Maximum state of dependence.
- Immobile,
- Respiratory system.
- Cardiopulmonary physiotherapy
- Changes in posture
- Homecare program

Dal Bello-Haas V, Kloos AD, Mitsumoto H. Physical therapy for a patient through six stages of amyotrophic lateral sclerosis. Phys Ther. 1998;78(12):1312-24.

FUNCTIONAL STAGE MODEL

Independent stage:

Motor ability is preserved, with the patient walking and performing normal daily activities. There is a slight reduction in muscle strength and susceptibility to fatigue. The main aims are to keep motor functioning stable for as long as possible, to avoid muscle retractions and joint deformities, to reeducate about posture and to give guidance on the use of orthoses.

Semi-independent stage:

Individuals present difficulty i performing daily activities and the use of wheelchairs is necessary. This is the start of respiratory system involvement, with dyspnea during moderate effort. Stretching, muscle strengthening, torso posture exercises and respiratory kinesiotherapy exercises are recommended. These procedures increase flexibility, reduce cramp, strengthen the musculature and improve the posture.

Dependent stage:

Patients require caregivers to assist them in performing day-to-day activities because of the evolution of the symptomatology. Preservation of joint mobility with emphasis on the pelvic and scapular regions, preservation or improvement in control over the torso and neck, respiratory training and postural changes are recommended

Piemonte MEP, Ramirez C. Como este manual pode ajudar. Piemonte MEP, editor. Manual de exercícios domiciliares para pacientes com esclerose lateral amiotrófica. São Paulo: Manole; 2001. pp. 19-64.

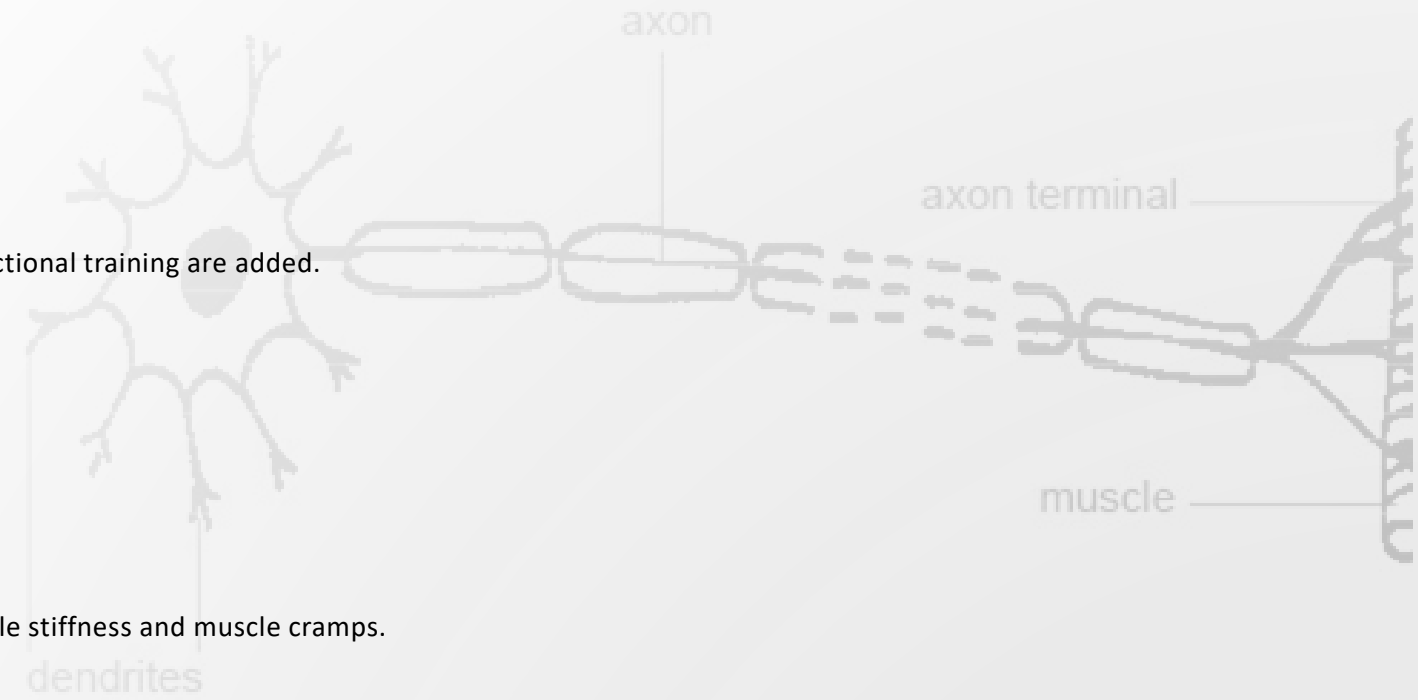
- An interesting stage progression model is one that focuses on when the patient wants involvement. This framework was developed with care advisors in mind and did not focus on physiotherapists, however it reminds us of the role of the physio to provide informed consent and to balance that keenly with patient and family autonomy.

Gilly Smith (in the Australasian Journal of neuroscience, Volume 25 ɣ Number 1 ɣ May 2015, Describing the role and function of Care Advisors in the Motor Neurone Disease Association of West Australia.)

Mrs. L and her husband were able acquire funding for 20 sessions of physio. They chose to use a handful spread out over a period of time.
They wanted to engage in life and family and not immerse themselves in the medical engagements more than they felt absolutely necessary.


INTERVENTIONS

INTERVENTION



- cardiopulmonary and neuromuscular conditioning and physical-functional training are added.
- **torso posture exercises**
- **posture guidance**
- Assistive and supportive device subscription.
- Education regarding how to find optimal positions
- stretching exercises: stretching can be a good intervention for muscle stiffness and muscle cramps.
- manual therapy: including mobs and massage.
- **Electrical stimulation**
- **hydrotherapy**
- Cardiopulmonary physiotherapy
- Position changing
- **homecare program**

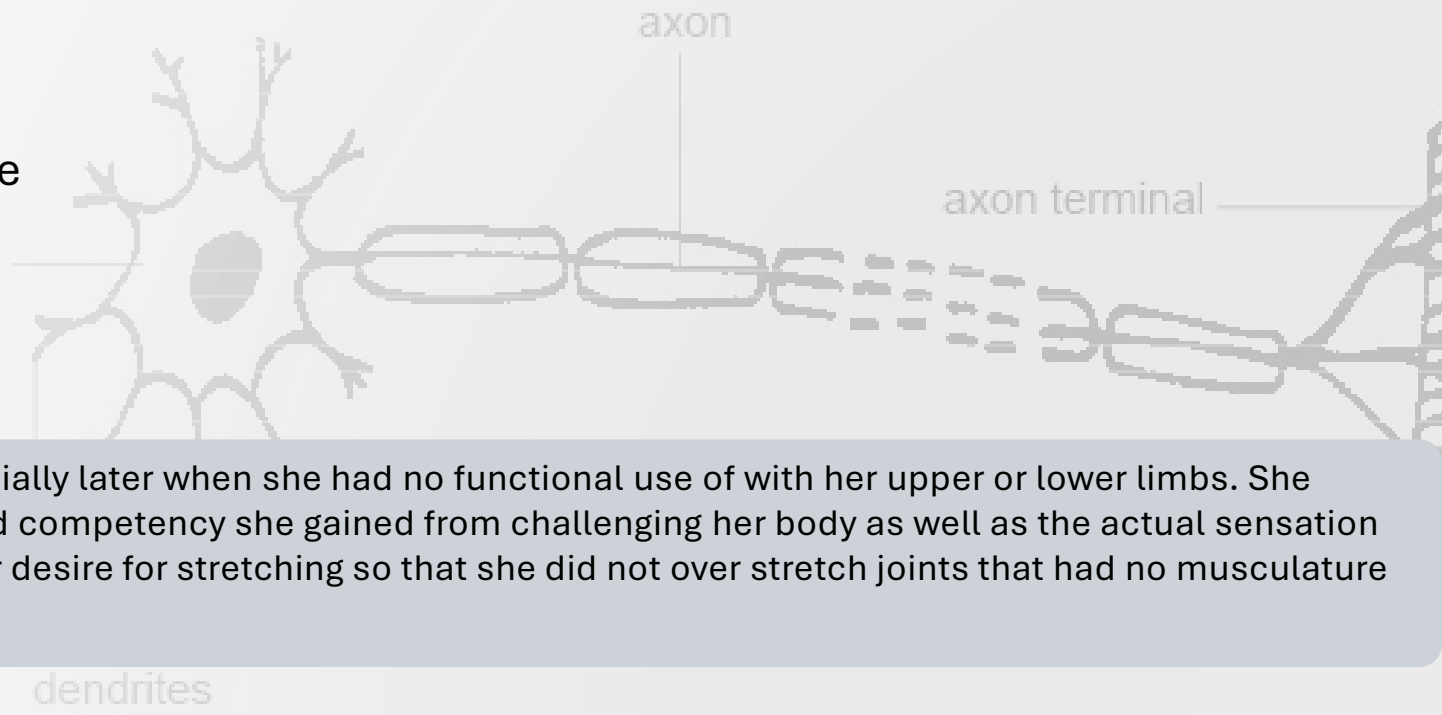
EARLY STAGE INTERVENTION

Assess and maintain muscle strength and function.

To improve mobility and flexibility; We have a number of modalities in our tool set here, we can use both strengthening and stretching exercises here as well as manual therapy and massage.

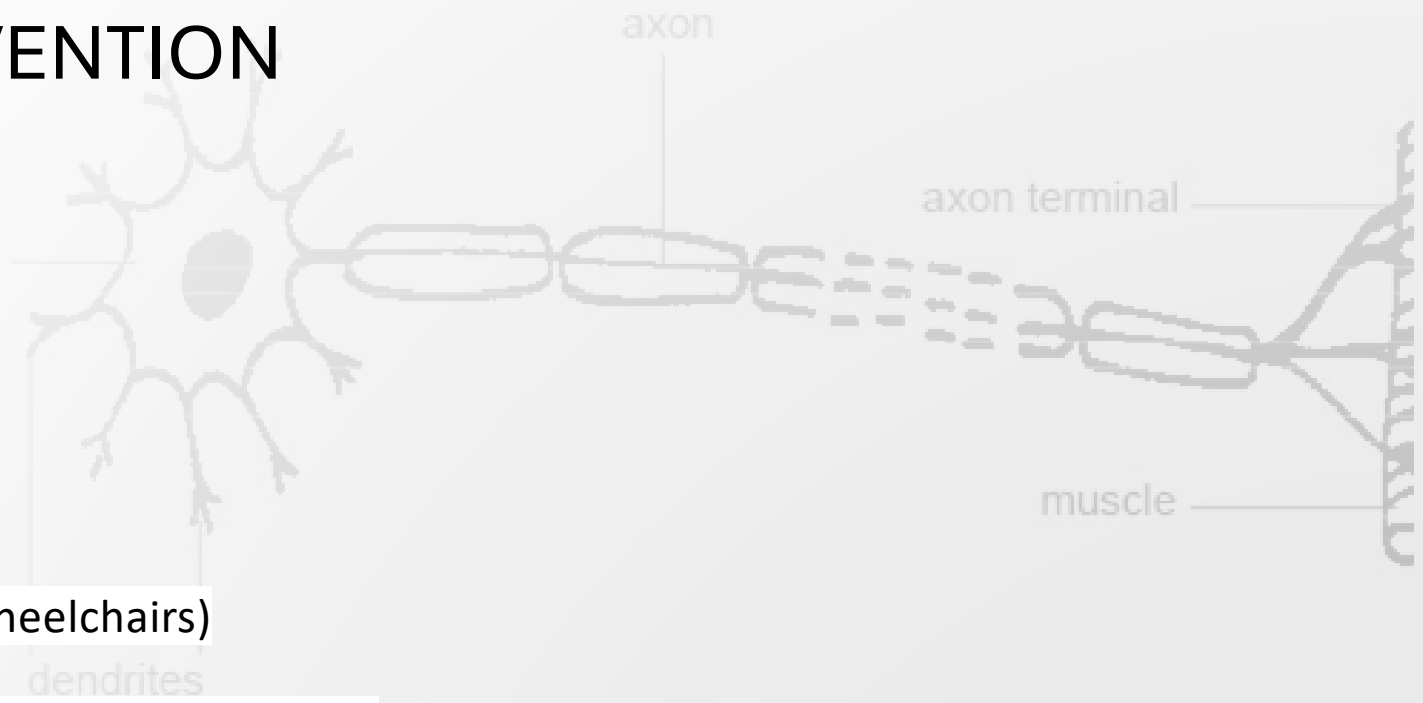
Enhancing balance and coordination.

Promoting functional activities and independence



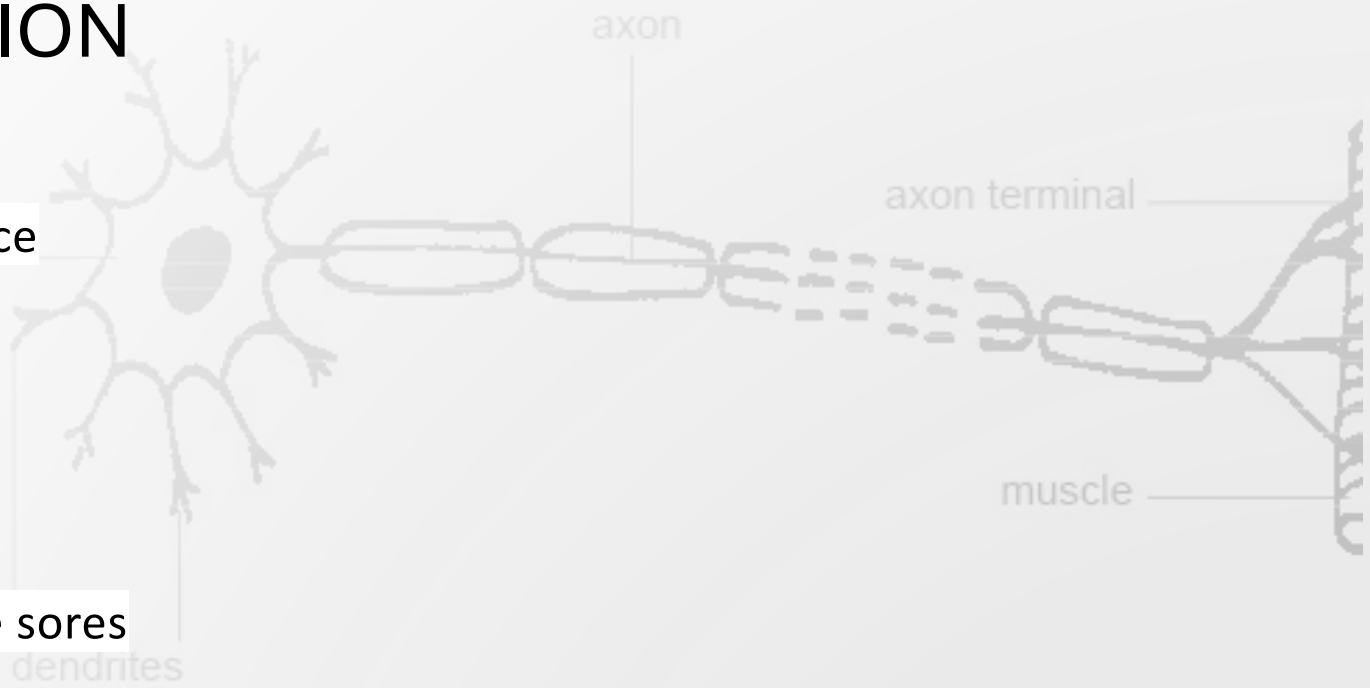
Mrs. C really enjoyed stretching. Especially later when she had no functional use of with her upper or lower limbs. She enjoyed the feeling of achievement and competency she gained from challenging her body as well as the actual sensation of stretching. I had to temper her eager desire for stretching so that she did not over stretch joints that had no musculature support.

PROGRESSIVE STAGE INTERVENTION



- Manage muscle weakness and atrophy
- Maintain range of motion and flexibility
- Assist with mobility aids (canes, walkers, wheelchairs)
- Develop strategies for daily activities and energy conservation
- Provide education on adaptive equipment and techniques

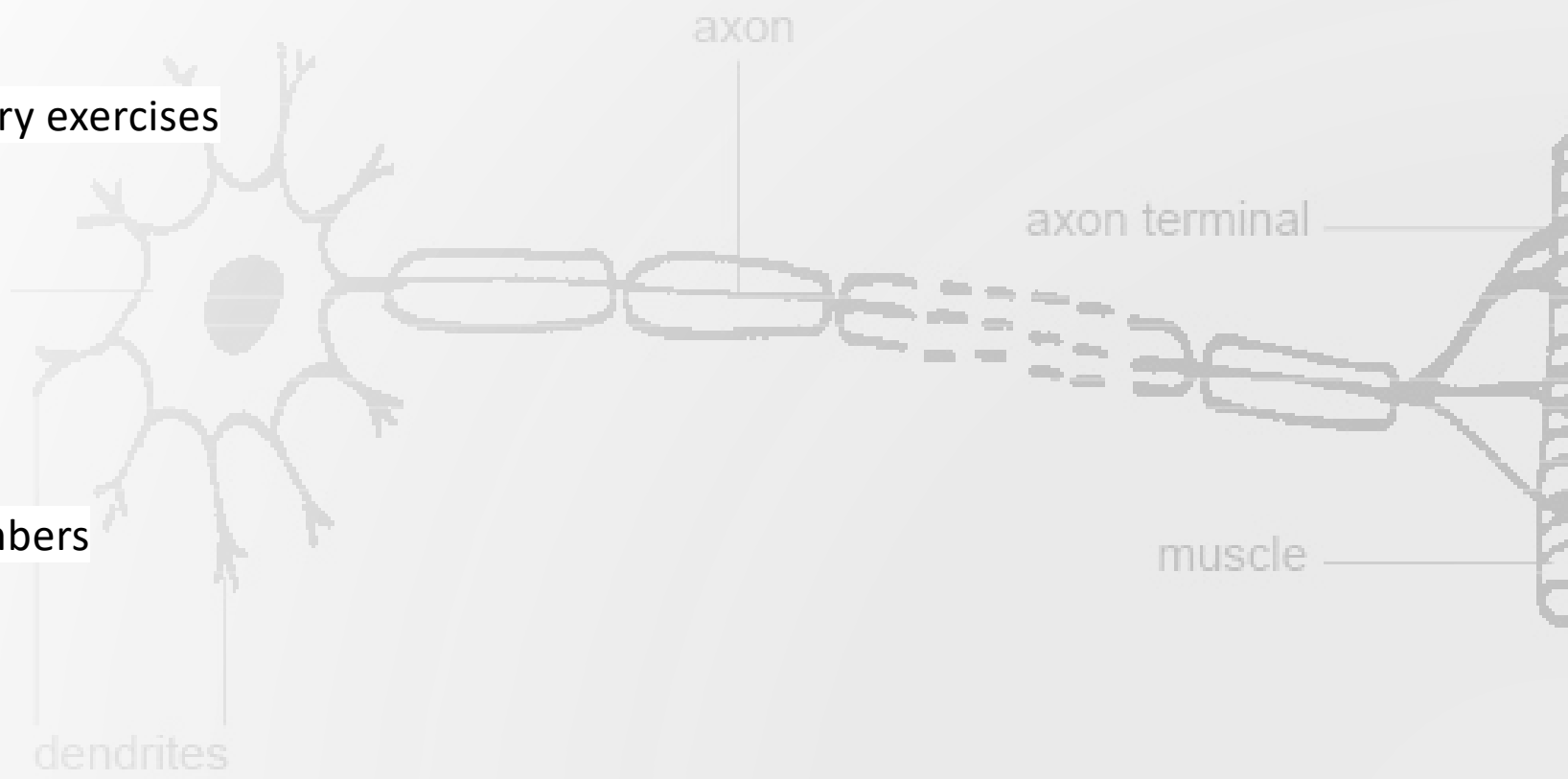
ADVANCED STAGE INTERVENTION



- Focus on respiratory and cardiac maintenance
- Manage pain and discomfort
- Maintain skin integrity and prevent pressure sores
- Assist with positioning and mobility techniques
- Provide emotional support and counseling

END STAGE INTERVENTION

- Provide palliative care and comfort measures
- Assist with breathing and respiratory exercises
- Maintain dignity and quality of life
- Support caregivers and family members



- Because we spend time with clients, we have a window of opportunity to build compassionate relationships with clients and to offer emotional support and counseling to patients and caregivers. Education, guidance and support must be given within our expertise and means and effective referral to other members of the health team is an essential aspect here. Psychologists, counselors, the MNDA care aids, and when necessary (if we notice the disease cognitive and or behavioral changes) neuro psychologists.

- It has become clear that ALS is more than a motor disease, there are sensory and cognitive implications.

- The non-pyramidal features of ALS result from the diesase's impact on the autonomic nervous system, the basal ganglia, and the cerebellar, frontotemporal, oculomotor, and sensory systems.

McCluskey, L Vandriel, S, Elman, L. (2014) 'ALS-Plus syndrome: non-pyramidal features in a large ALS cohort' *J Neurol Sci,* pp3451

Cited in Dal Bello-Haas, V. (2018) 'Physical therapy for individuals with amyotrophic lateral sclerosis: current insights', Degenerative Neurological and Neuromuscular Disease, Vol8, pp45-54. Available at: https://www.tandfonline.com/doi/full/10.2147/DNND.S146949

- Stage model: using hind and fore sight and plan for future intervention.
- **Activity limitations**
- **Participation**
- Symptoms and impairments
- Early intervention: breathing, posture, body movement, balance
- Early considerations can also help in preventing secondary complications such as pain and contractures.
- A fluid approach to the suggested intervention of what is described in advanced stage or semi independent stage can be introduced in the stage before.

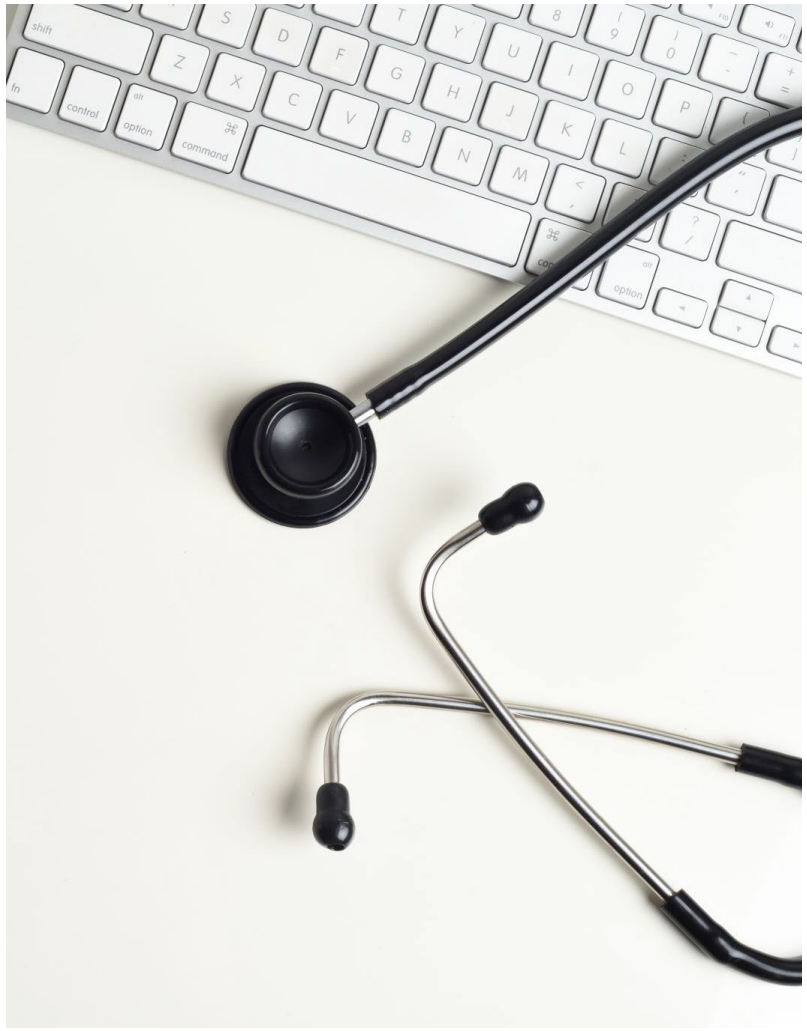
Early Stage Intervention Research

Gómez Fernández and Calzada Sierra (2001) used a rehabilitation program to maintain functional adaptation and prevent complications from muscle immobility.

Six patients with ALS underwent seven hours of rehabilitation per day, three times a week, for four weeks, while avoiding fatigue.

They used respiratory and motor kinesiotherapy, by means of stretching and muscle strengthening exercises, postural reeducation, facial mimicking exercises and body relaxation.

The results were significant: four of the six patients presented with improvement in their quality of life with early intervention.



Gómez Fernández L, Calzada Sierra DJ. (2001) 'Importancia del tratamiento rehabilitador multifactorial en la esclerosis lateral amiotrófica. [The importance of multifactorial rehabilitation treatment in amyotrophic lateral sclerosis]', *Rev Neurol*., Vol 32(5), pp. 423-6. Cited in Pozza, AM, Delamura, MK, Ramirez C, Valério NI, Marino LHC, Lamari NM. (2006) 'Physiotherapeutic conduct in amyotrophic lateral sclerosis'. *Sao Paulo Medical Journal*, Vol 124(6), pp. 350–4. Available from: https://www.scielo.br/j/spmj/a/WDyFZXFvnVswVsqxGpLcnMf/

Early Stage Intervention Research

Piemonte and Ramirez (2001) also concluded that early intervention by a multidisciplinary team offered improved quality of life and increased longevity.

Piemonte MEP, (2001), Manual de exercícios domiciliares para pacientes com esclerose lateral amiotrófica. São Paulo: Manole; 2001. p. 19-64.

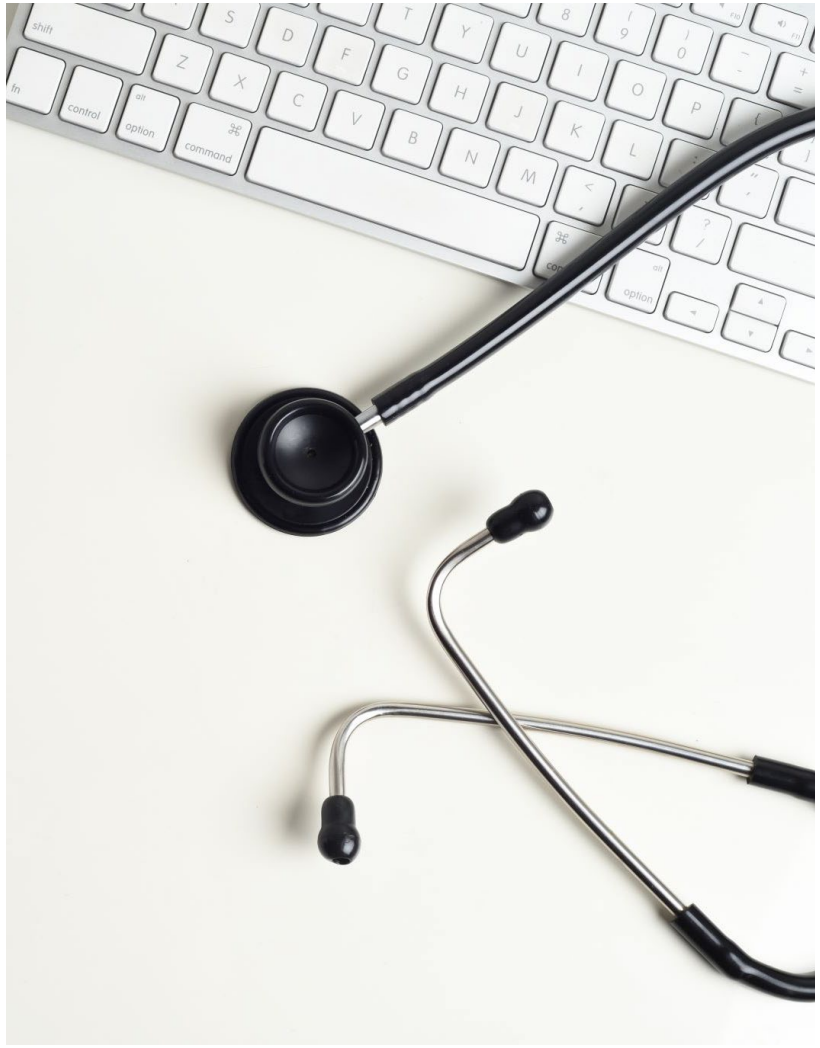
Cited in Pozza, AM, Delamura, MK, Ramirez C, Valério NI, Marino LHC, Lamari NM. (2006) 'Physiotherapeutic conduct in amyotrophic lateral sclerosis'. *Sao Paulo Medical Journal*, Vol 124(6), pp. 350–4. Available from: https://www.scielo.br/j/spmj/a/WDyFZXFvnVswVsqxGpLcnMf/

Early Stage Intervention Research

However, Pedroso et al. did not achieve the same results. They administered daily physiotherapy sessions to ALS patients in their homes over a six-month period, using motor and respiratory exercises to delay disease evolution and reduce the costs for the patients. There were two groups: independent and semi-independent patients. From these groups, a test group was formed composed of five independent and three semi-independent individuals who underwent functional, fatigue and muscle force evaluations. A control group was formed, composed of five independent and three semi-independent patients who did not perform these exercises. After evaluating the patients every three months over a one-year period, no significant differences were seen between the two groups.

Pedroso JPC, Ramirez C, Silva HCA, et al. Importance of the physical therapeutic program in muscular force and the functional state in patients with amyotrophic lateral sclerosis during 6 months. Proceedings of the 12th International Symposium on ALS/MND; 2001 Nov 18-20; Oakland, USA; 2001. p. 94.

Cited in Pozza, AM, Delamura, MK, Ramirez C, Valério NI, Marino LHC, Lamari NM. (2006) 'Physiotherapeutic conduct in amyotrophic lateral sclerosis'. *Sao Paulo Medical Journal*, Vol 124(6), pp. 350–4. Available from: https://www.scielo.br/j/spmj/a/WDyFZXFvnVswVsqxGpLcnMf



General Treatment Principles

When treating we have to decide which impairments, activity limitations, and participation restrictions:

1) can be restored;

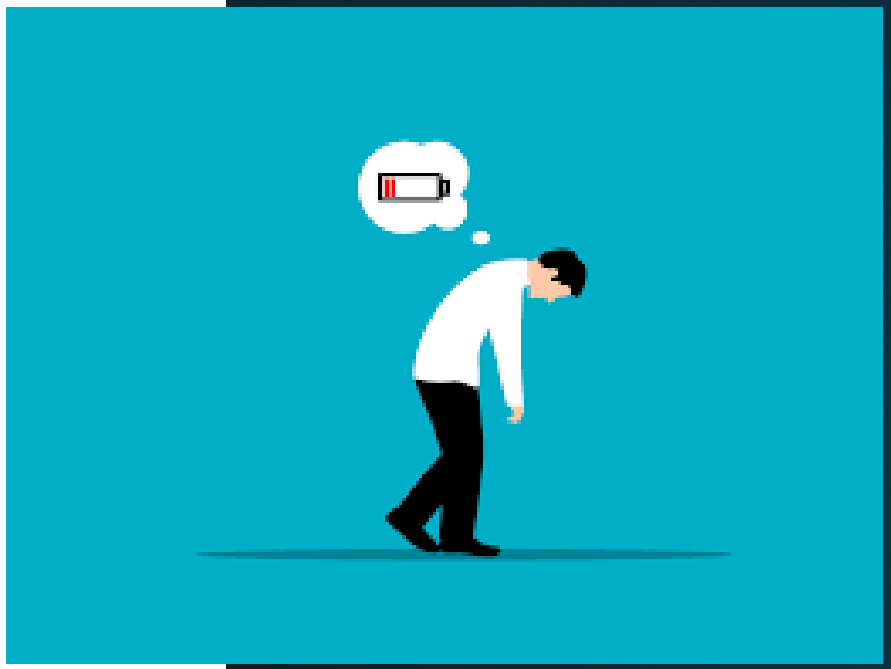
2) require compensatory strategies or interventions;

3) require referral to different health care professional(s); and

4) cannot be affected by physical therapy interventions at all.

FATIGUE

- 90% reported fatigue symptoms
- The least treated

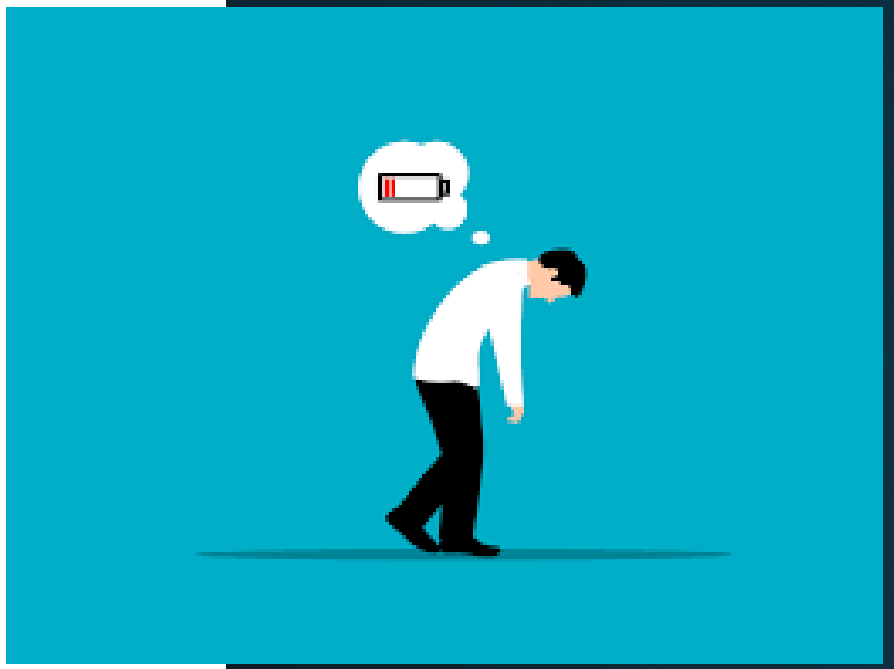


- NicholsonK MurphyA McdonnellE, Improving symptom management for people with amyotrophic lateral sclerosis Muscle and Nerve 2017 571202428561886)in Dal Bello-Haas

FATIGUE

Treatment

- Pacing
- Exercise moderation
- Efficient use of the body
- Relaxation and breathing techniques



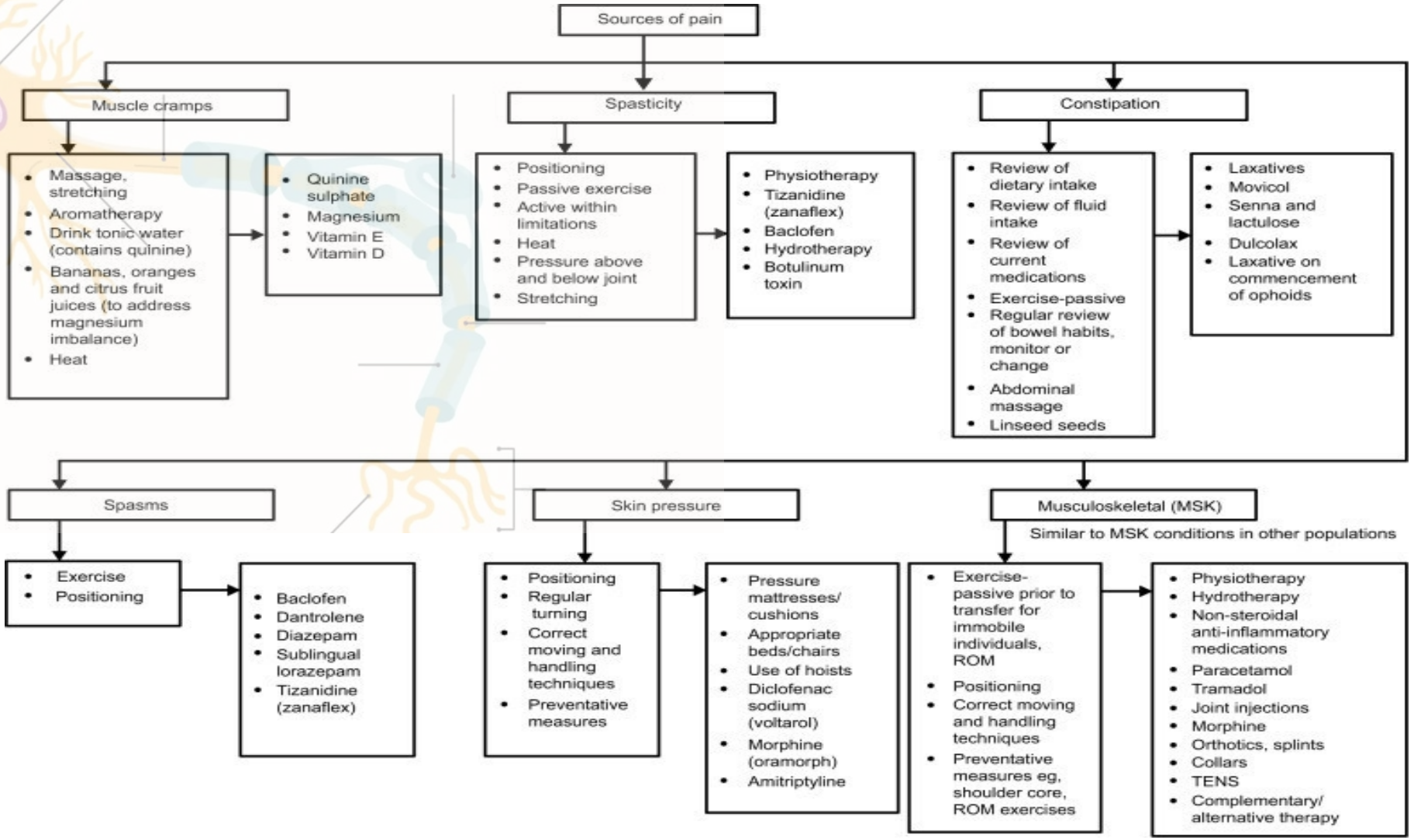
PAIN

Although ALS does not involve the pain pathways primarily

- Musculoskeletal impairments, immobility, loss of ROM, decreased support from weakened muscles, positioning difficulty, dependent edema, and acute injuries (sprains, strains, and falls)
- Spasticity and cramps, especially if severe, and pre-existing conditions

PAIN

- Range of motion (ROM) exercises,
- Passive stretching,
- Joint mobilizations, and
- Education about proper joint support and protection.



Dal Bello-Haas, V. (2018) "Physical therapy for individuals with amyotrophic lateral sclerosis: current insights", Degenerative Neurological Neuromuscular Disorders, Vol 8, pp. 45-54. Available at: 10.2147/DNND.S146949

FALLS

- Sanjak, M. et al. (2014) considered that 37% of ambulatory individuals with relatively normal clinical balance and mobility test findings (eg, Dynamic Gait Index, Berg Balance Scale Score, Timed Up and Go, Stair-Climbing Test, 25-Feet Walk Test)
- “had decreased ability to use vestibular input and required increased reliance on visual input for postural orientation to sustain equilibrium, as detected via sensory organization testing.“

Sanjak, M., Hirsch, M.A., Bravver, E.K., Bockenek, W.L., Norton, H.J. and Brooks, B.R., 2014. Vestibular deficits leading to disequilibrium and falls in ambulatory amyotrophic lateral sclerosis. *Archives of Physical Medicine and Rehabilitation*, *95*(10), pp.1933-1939.

- These authors suggested that peripheral and central pathologic abnormalities or ALS-related cerebellum pathology may have contributed to the vestibular deficits.

FALLS

- The involvement of the cerebellum in the pathologic abnormalities of ALS is increasingly recognized, lending further support that ALS is a multisystem, rather than purely a motor disease.
- PrellTGrosskreutzJThe involvement of the cerebellum in amyotrophic lateral sclerosisAmyotroph Lateral Scler Frontotemporal Degener2013147–850751523889583
- We need to ask about and assess recent falls at each visit. We can intervene with prescription of assistive devices or orthotics, and targeted balance training, and include vestibular training if appropriate.

- Dal Bello-Haas, V. (2018) 'Physical therapy for individuals with amyotrophic lateral sclerosis: current insights', *Degenerative Neurological and Neuromuscular Disease*, Vol 8, pp. 45-54. Available at https://www.tandfonline.com/doi/full/10.2147/DNND.S146949

EXERCISE

- Exercise, and fitness in general, remain **controversial** in the management of people with ALS.
- It is **often discouraged** because of concerns regarding overuse weakness and fatigue, as well as occasionally, a defeatist attitude (it won't really make any difference).
- However, the view that exercise, when prescribed appropriately, may be **physiologically and psychologically beneficial** for people with MND, especially when implemented in the earlier stages of the disease, is being increasing adopted.

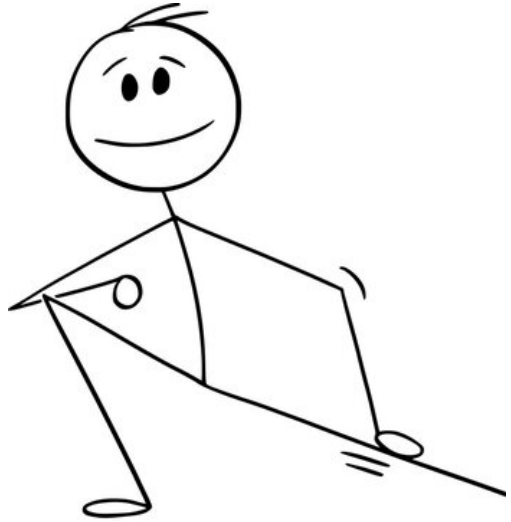
- Paganoni et al. Paganoni, S., Karak, C., Joyce, N.,Bedlack, R., Carter, G. (2015) 'Comprehensive Rehabilitative Care Across the Spectrum of Amyotrophic Lateral Sclerosis', *Neuro Rehabilitation*, 37(1), pp. 53-68.

EXERCISE

- A clinical trial with mice showed that moderate endurance exercise **delayed disease**

- Carreras et al., 2010; Kirkinezos, Hernandez, Bradley, and Moraes, 2003; Veldink et al., 2003) Cited in Dal Bello-Haas, V. (2018) 'Physical therapy for individuals with amyotrophic lateral sclerosis: current insights' Degenerative Neurological and neuromuscular Disease, Vol8, pp45-54. Available at: https://www.tandfonline.com/doi/full/10.2147/DNND.S146949

- In a small study with people with ALS, 25 people were randomized to perform an individualized moderate-intensity daily exercise program "involving most muscle groups of the four limbs and trunk", as opposed to avoiding any physical activity beyond their usual daily requirements. Drory et al., 2001). The **program was well tolerated** and was associated with less functional decline on the ALS this was also on the follow up after 3 months.



- Drory VE, Goltsman E, Reznik JG, Mosek A, Korczyn AD. The value of muscle exercise in patients with amyotrophic lateral sclerosis. J Neurol Sci. 2001;191(1–2):133–137. [PubMed] [Google Scholar] [Ref list] Cited in: Paganoni, S., Karak, C., Joyce, N.,Bedlack, R., Carter, G. (2015) 'Comprehensive Rehabilitative Care Across the Spectrum of Amyotrophic Lateral Sclerosis', Neuro Rehabilitation, 37(1), pp. 53-68.

- In a randomized, controlled trial Bello-Haas et al. (2007) showed that moderate resistance exercise in 27 people with ALS also resulted in **better function** at 6 months, as measured by total ALSFRS (ALS Functional Rating Scale) scores and quality of life, without adverse effects.

  Bello-Haas VD, Florence JM, Kloos AD, Scheirbecker J, Lopate G, Hayes SM, et al. A randomized controlled trial of resistance exercise in individuals with ALS. Neurology. 2007;68(23):2003–2007. [PubMed] [Google Scholar] [Ref list] Cited in: Paganoni, S., Karak, C., Joyce, N.,Bedlack, R., Carter, G. (2015) 'Comprehensive Rehabilitative Care Across the Spectrum of Amyotrophic Lateral Sclerosis', Neuro Rehabilitation, 37(1), pp. 53-68.

EXERCISE

- High intensity endurance exercise has proven to be detrimental; so we need to remain vigilant: muscle soreness, post exercise pain and muscle soreness which we usually embrace as a 'job well done' can indicate overwork induced muscle damage.

Petrof, BJ. (1998) 'The molecular basis of activity-induced muscle injury in Duchenne muscular dystrophy', Molecular and cellular biochemistry, 179(1–2), pp.111–123. Cited in: Paganoni, S., Karak, C., Joyce, N.,Bedlack, R., Carter, G. (2015) 'Comprehensive Rehabilitative Care Across the Spectrum of Amyotrophic Lateral Sclerosis', Neuro Rehabilitation, 37(1), pp. 53-68.

- We also need to balance the value of exercise with a frantic or desperate need to exercise (the family's, the person's, as well as the physio need to "do something")
- Something to keep in mind is that QOL does not directly correlate with strength and physical function

(Chio et al., 2004; Grehl, Rupp, Budde, Tegenthoff, & Fangerau, 2011; Simmons, Bremer, Robbins, Walsh, & Fischer, 2000). Cited in: Paganoni, S., Karak, C., Joyce, N.,Bedlack, R., Carter, G. (2015) 'Comprehensive Rehabilitative Care Across the Spectrum of Amyotrophic Lateral Sclerosis', Neuro Rehabilitation, 37(1), pp. 53-68.

- Rather, quality of life depends primarily on psychological and existential factors

(Bremer, Simone, Walsh, Simmons, & Felgoise, 2004; Calvo et al., 2011; Chio et al., 2004; Simmons et al., 2000). Cited in: Paganoni, S., Karak, C., Joyce, N.,Bedlack, R., Carter, G. (2015) 'Comprehensive Rehabilitative Care Across the Spectrum of Amyotrophic Lateral Sclerosis', Neuro Rehabilitation, 37(1), pp. 53-68.

Useful principles based on the above research

Do not exercise muscles that do not have antigravity strength

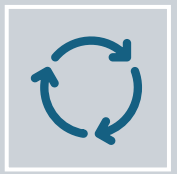
Avoid high resistance exercise

Avoid eccentric exercise

Useful principles based on the above research

Start early

Perform aerobic exercise at a moderate, sub-maximum level



Progress as tolerated (“start low, go slow”)

Gentle daily flexibility regime