Amyotrophic Lateral Sclerosis: Management Across the Disease Continuum

Goals and Objectives

Course Description

"Amyotrophic Lateral Sclerosis" is a live (real-time) interactive webinar for occupational therapists and occupational therapy assistants presenting contemporary knowledge on this progressive neurodegenerative disease. The course addresses pathophysiology, clinical presentation, and disease progression with emphasis on motor neuron loss, respiratory compromise, and bulbar involvement. Participants will learn strategies for managing functional decline and promoting quality of life across all stages of the disease.

Course Rationale

The purpose of this course is to enhance clinical understanding of the unique and evolving needs of individuals living with ALS. The evidence-based strategies presented will guide the development of individualized treatment programs that optimize independence and support functional preservation, safety, and quality of life.

Course Goals and Objectives

Upon completion of this course, participants will be able to:

1. Identify genetic mutations in familial and sporadic types of ALS.
2. Identify relevant biomarkers and imaging findings associated with diagnosis.
3. Recognize the stages of disease progression.
4. Identify assessment tools to track disease progression.
5. List the motor, non-motor, and bulbar components of clinical presentation.
6. Identify symptom management and psychosocial care strategies.
7. Distinguish the primary and secondary mechanisms contributing to pain.
8. Define respiratory compromise and recognize ventilation support options.
9. Identify dysphagia concerns and nutritional support strategies.
10. Identify contemporary options to facilitate communication.

Course Provider – Innovative Educational Services

Provider Contact Information – information@cheapceus.com

Course Instructor – Jodi Gootkin, PT, MEd, CEAS

Conflict of Interest – No financial or non-financial conflict of interest exists for the presenter or provider of this course.

Target Audience – Occupational Therapists, Occupational Therapy Assistants

Course Educational Level – This course is applicable for introductory learners.

Course Prerequisites – None

Method of Instruction – Live synchronous interactive webinar

Location - Cheapceus.com

Date – Multiple presentation dates.

Course Completion Requirements / Criteria for Issuance of CE Credits – Verified attendance at live interactive webinar and a score of 70% or greater on the course post-test

Continuing Education Credits – Three (3) contact hours / .3 AOTA CEUs

Course Fee - $39.95

Registration Information –To register for this program, please go to: Cheapceus.com

Special Needs Requests – Email: information@cheapceus.com or phone: 954-663-4101

Cancellation by the Learner – Learners may cancel their participation at any time and receive a full refund of all paid fees.

Cancellation by the Provider – Registrants will receive a full refund of all paid fees within 24 hours of provider cancellation.

Complaint Resolution – Please call 954-663-4101 (24 hours/day, 7 days/week) to speak with a live customer service agent. Our goal is to work with our customers to resolve all issues to the customer's satisfaction with just one phone call whenever possible.

Refund Policy - Unrestricted 100% refund upon request. The request for a refund by the learner shall be honored in full without penalty or other consideration of any kind. The request for a refund may be made by the learner at any time without limitations before, during, or after course participation.

American Occupational Therapy Association Approved Provider

Innovative Educational Services is an AOTA Approved Provider of professional development. PD activity approval ID# 5471. This Distance Learning – Interactive PD activity is offered at .3 CEUs; Introductory; Occupational Therapy Service Delivery; OT Foundational knowledge. The assignment of AOTA CEUs does not imply endorsement of specific course content, products, or clinical procedures by AOTA.

Amyotrophic Lateral Sclerosis: Management Across the Disease Continuum

Course Outline and Schedule

Topic	Time
Overview	0:00 - 0:10
Genetic and Environmental Factors	0:11 – 0:20
Pathogenesis	0:21 – 0:30
Neuroimaging and Biomarkers	0:31 – 0:40
Clinical Presentation	0:41 – 0:50
Interactive Discussion of Clinical Applications	0:51 - 1:00
Disease Progression	1:00 - 1:15
Motor Impairment	1:16 – 1:25
Primary and Secondary Pain	1:26 – 1:30
Cognitive and Behavioral Changes	1:31 - 1:40
Respiratory Compromise	1:41 – 1:50
Interactive Discussion of Clinical Applications	1:51 - 2:00
Dysphagia and Nutritional	2:00 – 2:15
Dysarthria and Communication	2:16 – 2:31
Pharmacotherapy	2:31 - 2:40
Patient Preferences and End of Life	2:41 - 2:50
Interactive Discussion of Clinical Applications	2:51 - 3:00

Amyotrophic Lateral Sclerosis: Management Across the Disease Continuum

Live Interactive Webinar
Presented By

Jodi Gootkin, PT, MED, CEAS
jodiemail@comcast.net

1

Course Overview

- "Amyotrophic Lateral Sclerosis" is a live (real-time) interactive webinar for rehabilitation professionals presenting contemporary knowledge on this progressive neurodegenerative disease. The course addresses pathophysiology, clinical presentation, and disease progression with emphasis on motor neuron loss, respiratory compromise, and bulbar involvement. Participants will learn strategies for managing functional decline and promoting quality of life across all stages of the disease.

2

2

Course Rationale

- The purpose of this course is to enhance clinical understanding of the unique and evolving needs of individuals living with ALS. The evidence-based strategies presented will guide the development of individualized treatment programs that optimize independence and support functional preservation, safety, and quality of life.

3

3

Goals and Objectives

1. Identify genetic mutations in familial and sporadic types of ALS.
2. Identify relevant biomarkers and imaging findings associated with diagnosis.
3. Recognize the stages of disease progression.
4. Identify assessment tools to track disease progression.
5. List the motor, non-motor, and bulbar components of clinical presentation.
6. Identify symptom management and psychosocial care strategies.
7. Distinguish the primary and secondary mechanisms contributing to pain.
8. Define respiratory compromise and recognize ventilation support options.
9. Identify dysphagia concerns and nutritional support strategies.
10. Identify contemporary options to facilitate communication.

4

4

Disclaimer

- Application of concepts presented in this webinar is at the discretion of the individual participant in accordance with federal, state, and professional regulations.
- No conflict of interest exists for the presenter or provider of this course.

5

5

Course Outline and Schedule

3-Hour
Live Interactive
Webinar

Topic	Time
Overview	0:00 - 0:15
Genetic and Environmental Factors	0:16 – 0:30
Pathogenesis	0:31 – 0:40
Neuroimaging and Biomarkers	0:41 – 0:50
Interactive Discussion of Clinical Applications	**0:51 - 1:00**
Disease Progression	1:00 - 1:20
Motor Impairment	1:21 – 1:50
Interactive Discussion of Clinical Applications	**1:51 - 2:00**
Respiratory Compromise	2:00 – 2:15
Bulbar Involvement, Nutrition, and Communication	2:16 – 2:31
Fatigue, Pain, Cognitive and Behavioral Changes	2:31 - 2:40
Pharmacotherapy	2:41 - 2:45
Patient Preferences and End of Life	2:46 – 2:50
Interactive Discussion of Clinical Applications	**2:51 - 3:00**

6

6

How To Obtain CEUs For This Course

- Course review and summary for post test at the end of the webinar.
- After the live interactive webinar and prior to 11:59 pm TONIGHT go to cheapceus.com
- Complete the post test with score of at least 70%
  - May be retaken multiple times
- Submit online payment for course
- Print certificate

7

7

Course Post Test

- Slides with "Consider This" icon in bottom right corner will be helpful in completing the post-test.

Consider This

8

8

Overview

- A rare and progressive neurodegenerative disorder of the central and peripheral nervous system leading to advancing weakness, loss of voluntary movement, and ultimately life-limiting respiratory decline.
  - Amyotrophic Lateral Sclerosis (ALS)
  - Lou Gehrig's Disease

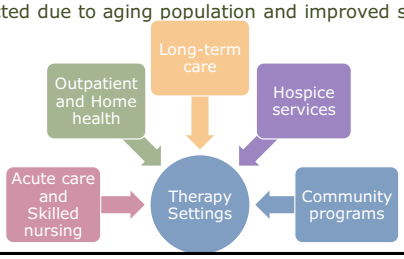
Consider This

9

9

Incidence and Prevalence

- Steady increase in people living with ALS world-wide is expected due to aging population and improved survival.

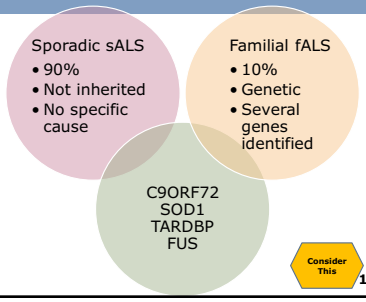


10

10

Genetics

- Etiology not fully understood, likely involves genetic susceptibility and environmental influences.



Consider This

11

11

Environmental Risk Factors

- Associations correlate with epigenetic alterations, but direct causality is not established.
- DNA methylation silences genes essential for synaptic integrity, axonal transport, and neuronal viability.

Newell, M. E., et.al. (2025). Epigenetic Biomarkers Driven by Environmental Toxins Associated with Alzheimer's Disease, Parkinson's Disease, and Amyotrophic Lateral Sclerosis in the United States: A Systematic Review. *Toxics*, *13*(2), 114. Benatar, M., Heiman-Patterson, T. D., Cooper-Knock, J., Brickman, D., et.al. (2025). Guidance for clinical management of pathogenic variant carriers at elevated genetic risk for ALS/FTD. *Journal of Neurology, Neurosurgery & Psychiatry*, 96(3), 209-218.

12

12

Head Injury (HI) and Earlier Age of ALS Diagnosis

- Not an absolute risk factor or cause.

Associated with Diagnosis before age 60

- Childhood or early adulthood HI
- Hospital visit or loss of consciousness
- Five or more HIs

Reverse Causation

- Mid to late adult HI and CTE
- Short interval between injury and diagnosis
- Prodromal ALS

Consider This

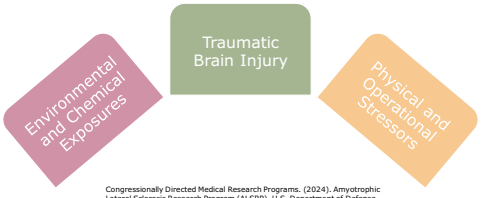
Raymond, J., et.al. (2025). Head Injury and Amyotrophic Lateral Sclerosis: Population-Based Study from the National ALS Registry. Brain Sciences, 15(2), 143.

13

13

Presumption of Military Service Connection

- Some occupational and environmental deployment exposures are associated with increased ALS risk, but no single exposure has been definitively proven as causal.



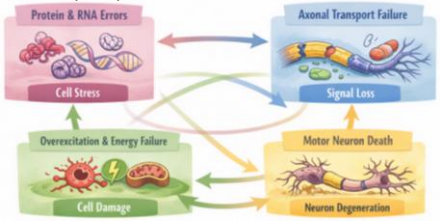
Congressionally Directed Medical Research Programs. (2024). Amyotrophic Lateral Sclerosis Research Program (ALSRP). U.S. Department of Defense.

14

14

Pathogenesis

- Mechanisms develop in parallel and reinforce one another over time.



Faller, K. M., et.al. (2025). Targeting common disease pathomechanisms to treat amyotrophic lateral sclerosis. *Nature Reviews Neurology*, 1-17. González-Sánchez, M., et.al. (2025). Pathophysiology, Clinical Heterogeneity, and Therapeutic Advances in Amyotrophic Lateral Sclerosis: A Comprehensive Review of Molecular Mechanisms, Diagnostic Challenges, and Multidisciplinary Management Strategies. *Life*, *15*(4), 647.

15

15

Cell Stress and Toxic Buildup

Key Factors

- Misfolded proteins (TDP-43)
- RNA processing errors

Consequences

- Proteins in the wrong place
- Disrupted cell functions
- Neuron becomes unstable
- Sets the stage for neurodegeneration

16

16

Signal Loss

Key Factors

- Axonal transport failure
- Cytoskeleton breakdown
- Blocked movement of cellular materials

Consequences

- Neurotransmitter depletion
- Mitochondrial dysfunction
- Axon retraction
- Loss of synaptic connections

17

17

Cell Damage

Key Factors

- Neurotransmitter imbalance
- Glutamate excitotoxicity
- Calcium overload
- Mitochondrial stress
- Diminished astrocyte clearance

Consequences

- Reduced neuronal efficiency
- Energy failure
- Inflammation
- Slower recovery

18

18

Neuron Degeneration

Key Factors

- Persistent calcium stress
- Chronic neurotransmitter dysregulation
- Sustained glial and astrocyte activation
- Failure of cellular repair
- Ongoing signal loss

Consequences

- Motor neuron death
- Ongoing muscle denervation
- Progressive weakness
- Declining functional capacity

19

19

Upper and Motor Neuron Involvement

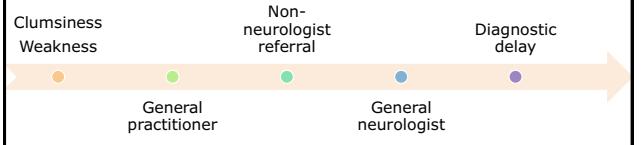
- Disrupts the motor network linking the primary motor cortex, lateral corticospinal tracts, anterior horn cells, and neuromuscular junctions in voluntary muscles.
- Brainstem involvement of bulbar motor cranial nerve nuclei.

20

20

Diagnosis Challenges

- Variable presentation and late symptom emergence lead to delayed diagnosis.
- No single diagnostic tool.

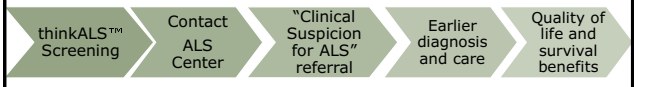


21

21

thinkALS™ tool

- This tool cannot be substituted as a diagnostic instrument but used only as a diagnostic guide.



Could this be ALS?
PROGRESSIVE and ASYMMETRIC MUSCLE WEAKNESS without radicular pain or sensory loss.

ALS Association. thinkALS™ for Faster Diagnosis. https://www.als.org/thinkals

22

22

Neuroimaging and Biomarkers

- No single confirmatory test
- Clinical criteria (El Escorial, Awaji, Gold Coast) identify a consistent pattern of progressive upper and lower motor neuron involvement.
- Electromyography (EMG)
- Magnetic Resonance Imaging (MRI)
- Positron Emission Tomography (PET)
- Diffusion Tensor Imaging (DTI)
- Fluid biomarkers, including neurofilament light chain (NfL)

García-Casanova, P. H., & Vázquez-Costa, J. F. (2025). Advances in the early diagnosis of amyotrophic lateral sclerosis. Expert Review of Neurotherapeutics, 25(4), 415-425.

23

23

Diffusion Tensor Imaging (DTI)

- A specialized type of MRI that measures how water molecules move inside brain tissue, especially along white-matter nerve fibers.
- Provides information about microstructural damage earlier than standard imaging.
  - Decreased Fractional Anisotropy (FA)
  - Increased Mean Diffusivity (MD)
  - Confirms UMN involvement

Qin, J., Wang, X., Fan, G., Zhang, W., Wu, X., Wang, B., & Liu, Y. (2024). Identifying amyotrophic lateral sclerosis using diffusion tensor imaging, and correlation with neurofilament markers. Scientific reports, 14(1), 28110.

Consider This

24

24

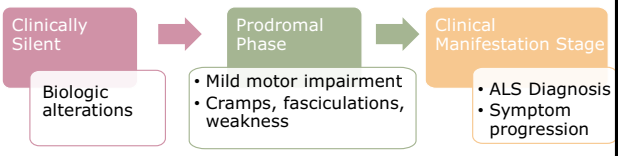
Neurofilament Light Chain (NfL) Levels

- NfL is a structural axonal protein that is released with motor neuron injury.
- Elevated serum and cerebrospinal fluid levels are indicative of disease severity and progression.
- Can support early diagnosis in high-risk individuals.

25

25

Disease Onset



26

26

Symptom Onset Patterns

- Variable early clinical signs
- Limb Onset
  - Asymmetrical upper or lower extremity weakness
  - Muscle fasciculations, decreased dexterity
- Bulbar Onset
  - Speech and/or difficulties, hoarse voice
- Respiratory Onset

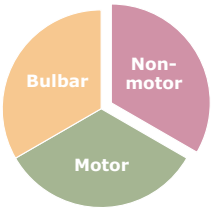
Consider This

27

27

Multisystem Disorder

- Sensation and eye muscle control is typically retained.



Pain
Fatigue
Behavior
Cognition
Constipation
Loss of appetite
Sleep disturbance
Urinary Incontinence

28

28

Disease Progression

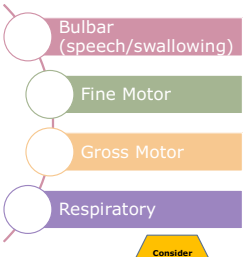
- No cure with treatment goals is to delay disease progression and manage symptoms as they emerge.

29

29

ALS Functional Rating Scale (ALSFRS-R)

- Measures functional decline across several domains.
- Higher scores indicate better function with lower scores reflecting greater disability.



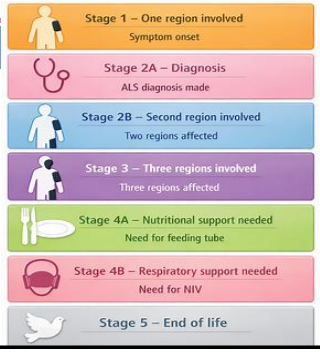
Consider This

30

30

King's Staging System

- Tracks disease spread



31

31

Milano-Torino Staging (MiToS)

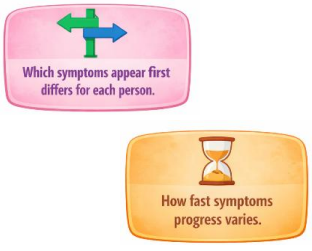
- Tracks functional decline

32

32

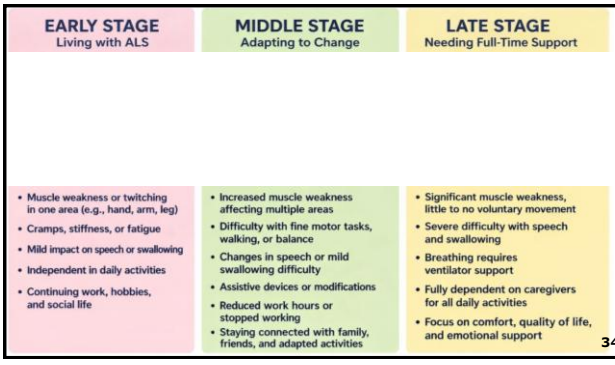
Disease Progression Milestones

- Employment change
- Mobility device
- Caregiver support
- Wheelchair use
- Communication aid
- Eye gaze technology
- Respiratory support
- Artificial nutrition
- Hospice



33

33



34

34

Motor Impairment

Variable and progressive spreading pattern of symptoms.

Initial site→ Adjacent area→ Contralateral side→ Proximal/distal spread

Upper Motor Neuron
- Voluntary muscle weakness
- Spasticity
- Hyperreflexia
- Loss of dexterity
- Pathological reflexes
  - Babinski, Hoffman's

Lower Motor Neuron
- Voluntary muscle weakness
- Fasciculations
- Atrophy

35

35

Strength – Atrophy Gap

- Denervation causes immediate weakness before atrophy appears.
- Reinnervation delays visible wasting until it collapses.
- Fast-twitch fibers fail first while slow fibers preserve bulk.
- Use strength testing and functional assessments vs. circumferential measurements to evaluate deficits.

36

36

Split Hand Index

- Disproportionate wasting of thenar and first dorsal interosseous muscles with relative sparing of hypothenar muscles.
  - Abductor pollicis brevis (APB)
  - First dorsal interosseous (FDI)
- Loss of precision/pincer grip.
  - Avoid high load isolation exercises.
  - Use lateral or palmar grasp, two hand strategies, thumb opposition support splinting and adaptive equipment.

37

37

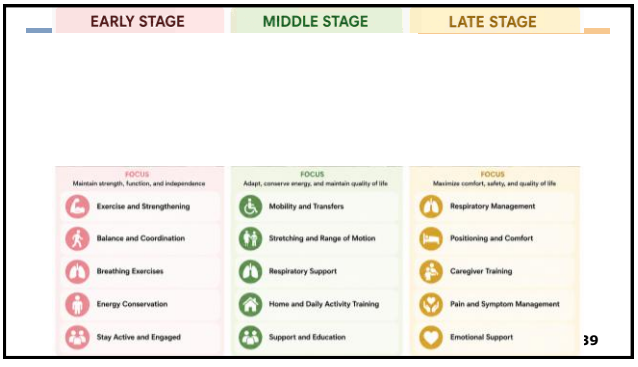
Motor Impairment Interventions

- Stretching and orthotics
- Balance training and fall assessment
- Activity modification
- Energy conservation and adaptive equipment
- Home modifications and emergency egress
- Caregiver training and social engagement
- Mobility aids, transfer equipment, wheelchair customization

Gratzer, A., et.al. (2025). Rehabilitation in Amyotrophic Lateral Sclerosis: Recommendations for Clinical Practice and Further Research. Journal of Clinical Medicine, 14(23), 8590.

38

38



39

39

Exercise Guidelines

- Moderate resistance and aerobic training
- Appropriate for muscles that can move against gravity.

Li, Z., & Kang, H. (2024). Efficacy of non-pharmacological interventions for individuals with amyotrophic lateral sclerosis: Systematic review and network meta-analysis of randomized control trials. Scientific reports, 14(1), 11365.

40

40

Exercise Outcomes

Intervention	Outcome
Animal study treadmill	High-intensity consistently worsened outcomes. Moderate delayed onset or improved survival in some cases. Swimming most consistent benefit.
Human resistance training	Beneficial effects on strength, function, quality of life. No evidence of slowing disease progression.
Human aerobic training	Beneficial effects on endurance, gait, work capacity, quality of life.

Fenili, G., et.al. (2024). Physical exercise in amyotrophic lateral sclerosis: a potential co-adjuvant therapeutic option to counteract disease progression. Frontiers in Cell and Developmental Biology, 12, 1421566.
Ren, S., et.al. (2025). The effect of exercise intervention on amyotrophic lateral sclerosis: A systematic review and meta-analysis. Frontiers in Neurology, 16, 1499407.

41

41

Targeted Regions

- Bracing for foot drop, wrist/hand weakness, neck extensor weakness and shoulder subluxation
- Stretching and positioning for shoulder girdle, hamstring and gastrocnemius tightness and/or spasticity
- Massage and stretching for cramps

42

42

Adaptive Equipment

- Bendable utensils, nosey cup, long flexible straws, one way straw, universal cuff, mobile arm support
- Bath mitt, hairdryer stand, grooming station, portable showers
- Suspenders, zipper shoes
- Leisure and pleasure activity adaptations

43

43

Mobility Equipment

- Cane, rollator walker
- Transfer vest, lift belt
- Gliding transfer boards, pivot disks, sliding bed sheets, swivel seat cushion, mechanical lift, track lift
- Raised seat, seat lift cushion, sit to stand lift
- Seat-lift recliner

44

44

Wheelchair Features

- Power tilt, power recline, elevating height power wheelchair
- Power articulating leg rests
- Vent tray
- Drive control -joystick, sip & puff, switch, chin, head-array, foot pedal, eye gaze, infrared, attendant control
- Seating system
- Home modifications

45

45

Integumentary Care

- Patient and family training in skin inspection, pressure relief positioning, deep vein thrombosis monitoring.
- Edema positioning, compression garments and massage

Sensation assessment

Elevate entire leg or arm above heart

Manage tight gastroc and hamstring

Tilt & recline wheelchair with articulating leg rests

Seat lift recliner

Sequential compression device (SCD)

46

46

Bowel and Bladder

- Constipation due to non-neurological factors
  - Managed with medication, diet, and exercise
- Urinary urgency, incontinence, and retention
  - Managed with medication, pelvic floor strengthening, and catheter
- Mobility, adaptive equipment, and sleep disturbance considerations
  - Lift commode, magnetic/velcro clothing closures, split seams, female urinal, wireless doorbell with gumball/buddy button

47

47

Respiratory Involvement

- Assess respiratory insufficiency.
  - Dyspnea, fatigue, yawning, orthopnea, accessory muscle use, cough strength, morning headaches, daytime sleepiness
- Nocturnal hypoventilation and obstructive sleep apnea
  - Modify sleep position and contributors to fragmented sleep
- Pulmonary complications and weakness become life limiting.

Macpherson, C. E., et.all. (2026). Physical Therapist Interventions for People With Amyotrophic Lateral Sclerosis Across Disease Stages: A Systematic Review of Efficacy. Physical Therapy, 106(1), pzaf142.

48

48

Respiratory Training

- Forced Vital Capacity (FVC) is a predictor of survival.
- Maximum Inspiratory Pressure (MIP) and Sniff Nasal Inspiratory Pressure (SNIP) measure muscle strength.
- Goal of interventions is to delay progression to ventilatory failure.
- Breathing exercises
- Inspiratory muscle training
- Posture training and bracing
- Airway clearance

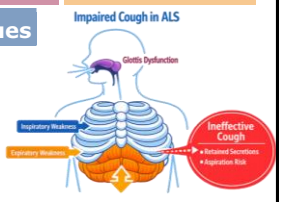
Consider This

Benzo-Iglesias, M. J., et.al. (2025). Efficacy of respiratory muscle training in improving pulmonary function and survival in patients with amyotrophic lateral sclerosis: a systematic review and meta-analysis. Therapeutic advances in respiratory disease, 19, 17534666251346095.

49

Airway Clearance Techniques

- Peak Cough Flow (PCF) measures cough strength.
- Manually assisted cough
- Mechanical insufflation/exsufflation
- Postural drainage
- Expiratory muscle training
- Portable suction
- Oral care

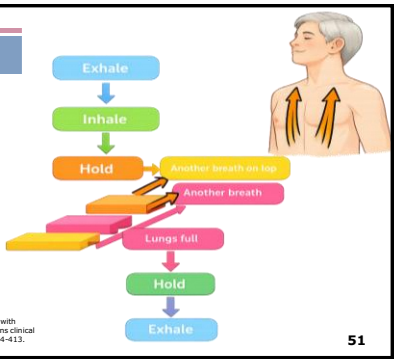


Magni, E., et.al. (2024). Effects of Respiratory Training on Pulmonary Function, Cough, and Functional Independence in Patients with Amyotrophic Lateral Sclerosis. Neurology International, 16(6), 1332-1342.
McHenry, K. L. (2024). Airway clearance strategies and secretion management in amyotrophic lateral sclerosis. Respiratory Care, 69(2), 227-237.

50

Breath Stacking

- Assists with oxygenation, secretion clearance, and thoracic mobility.



Khan, A., et.al. (2023). Respiratory management of patients with neuromuscular weakness: American college of chest physicians clinical practice guideline and expert panel report. Chest, 164(2), 394-413.

51

Lung Volume Recruitment (LVR)

- Incorporates manual bagging during inspirations of breath stacking.
  - Establish patient signal gesture of full lungs to STOP inflation.
  - Do NOT use bag for resuscitation emergencies.

Khan, A., et.al. (2023). Respiratory management of patients with neuromuscular weakness: American college of chest physicians clinical practice guideline and expert panel report. Chest, 164(2), 394-413.

52

Respiratory Outcomes

Study	Outcome
Fatigue prevalence	Higher fatigue frequency with ALSFRS-R $\leq$ 30. Correlated to lower functional status and poorer quality of life.
Respiratory muscle training	MEP and swallow function most consistent improvement. MIP inconsistent improvement. ALSFRS-R and FVC no significant improvement.

Hamad, A. A., et.al. (2024). Prevalence and correlates of fatigue in amyotrophic lateral sclerosis: A systematic review and meta-analysis. Neurological Sciences, 45(2), 485-493.
Ansari, U., et.al.(2025). Analysis of Respiratory Muscle Strength Training in Amyotrophic Lateral Sclerosis (ALS) Patients: A Systematic Review. *Cureus*, *17*(2).
Benzo-Iglesias, M. J., et.al. (2025). Efficacy of respiratory muscle training in improving pulmonary function and survival in patients with amyotrophic lateral sclerosis: a systematic review and meta-analysis. *Therapeutic advances in respiratory disease*, *19*, 17534666251346095.

53

Ventilation Support

- Noninvasive ventilation(NIV)
  - Bi-level Positive Airway Pressure (BiPAP)
  - Home ventilator
- Invasive ventilation
  - Tracheostomy

54

Bulbar Involvement

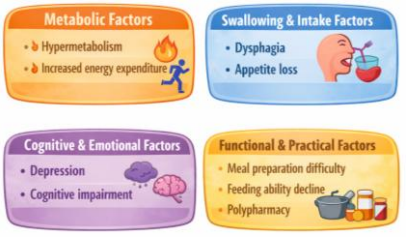
- Facial muscle weakness and temporal muscle wasting
- Palmomental Reflex
  - Thenar eminence stroke elicits chin twitch
- Jaw Jerk Reflex
  - Tapping chin elicits jaw clonus
- Sialorrhea

55

55

Malnutrition

- Weight stability is a prognostic factor for overall survival.



Bjelica, B., et.al. (2024). Non-motor symptoms in patients with amyotrophic lateral sclerosis: current state and future directions. *Journal of neurology*, *271*(7), 3953-3977.

56

56

Dysphagia

- Early screening minimizes risk of pneumonia due to aspiration.
- EAT-10 Screening Tool
- Assess bathroom accessibility
- Posture and expiratory muscle training
- Oral hygiene

Consider This

57

57

Diet and Eating Modifications

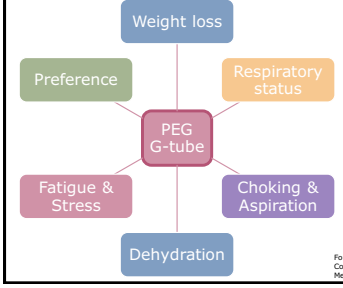
- Texture modified diet
- Eating efficiency
  - Meal preparation, feeding technique, and mealtime length
- Safe feeding techniques
  - Swallowing maneuvers
  - Breath stacking
  - Adaptive equipment
- Alleviate stress on diaphragm after eating

de Carvalho Vilar, M. D., et.al. (2025). Evidence-Based Nutritional Recommendations for Maintaining or Restoring Nutritional Status in Patients with Amyotrophic Lateral Sclerosis: A Systematic Review. Nutrients, 17(5), 782.

58

58

Nutritional Support



- Hypercaloric nutrition to maintain nutritional status, support body weight, and mitigate catabolic muscle loss leading to muscle wasting.
- Artificial nutrition

Consider This

Fouad, A. I., et.al. (2025). Evolution of Wheelchair Technology: A Comprehensive Overview of History, Disabilities, Types and Control Mechanisms. Advanced Sciences and Technology Journal, 2(2), 1-20.

59

59

Dysarthria

- Muscle spasticity and weakness
- Hypernasality
- Compensatory speech strategies
- Palatal lift
- Lung function and respiratory muscle strength
  - Expiratory muscle training
  - Lung volume recruitment

Pasqualucci, E., et.al. (2025). Management of dysarthria in amyotrophic lateral sclerosis. Cells, 14(14), 1048.

60

60

Alternative and Augmentative Communication

- Communication board
- Alerting system
- Voice amplifier, microphone
- Eye-gaze technology
- Speech synthesizers
- Message banking
- Voice banking

61

61

Non-Motor Symptoms

- Sleep disturbance
- Autonomic function
- Fatigue
- Pain
- Cognition
- Behavior

62

62

Fatigue

- Contributors to fatigue should be identified and managed.
  - Respiratory insufficiency, disrupted sleep, insufficient nutrition, prolonged mealtime, muscle weakness, depression, medication side effects
- Screen for comorbidities
- Prioritize energy conservation, positioning and mobility efficiency.

63

63

Pain

- Common later stage non-motor symptom that may be difficult for patient to report.

Primary Neuropathic	Secondary Mechanical **More Prevalent**
Burning, tingling Dysesthesia	Joint and soft tissue pain
Sleep disturbance	Limits mobility Sleep Disturbance

64

64

Pain Management

- Hydration, electrolytes and bowel regimen
- **Early Stage**
  - Postural correction, ergonomic adjustments and activity pacing
  - Modalities and soft-tissue techniques
- **Middle Stage**
  - Orthotics, bracing, and contracture prevention
  - Positioning schedule, sleep hygiene and seating optimization
- **Late Stage**
  - Pressure relief, comfort positioning and palliative coordination

65

65

Cognitive Changes

- Varying degrees of cognitive impairment to executive functioning.
  - Reduced verbal fluency
  - Mental inflexibility
  - Disinhibition
  - Impaired planning
- Some patients develop Frontotemporal Dementia (FTD).

66

66

Behavioral Changes

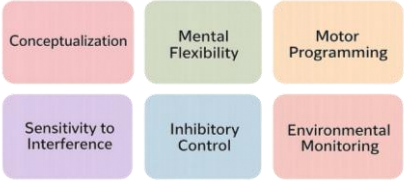
- Pseudobulbar affect
- Apathy, disinhibition, impaired social cognition, and depression

67

67

Frontal Assessment Battery (FAB)

- Screens for frontal lobe executive dysfunction abilities.
- Requires spared verbal and motor abilities for completion.



Aiello, E. N., et.al.(2023). Feasibility and diagnostics of the Frontal Assessment Battery (FAB) in amyotrophic lateral sclerosis. Neurological Sciences, 44(2), 587-592.

Consider This

68

68

ALS Specific Tools

Edinburgh Cognitive and Behavioral ALS Screen (ECAS)

- Executive functions, memory, language, visuospatial skills and social cognition
- Accounts for communication limitations to gather responses.

ALS Cognitive Behavioral Screen (ALS-CBS)

- Cognition assessment
  - Attention, concentration, mental tracking, and verbal fluency
- Behavioral interview with caregiver
  - Empathy, personality, judgment, language, and insight

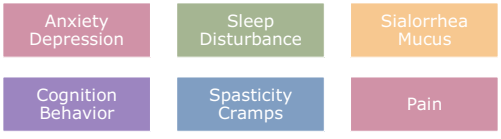
Didcote, L., et.al. (2024). What is the extent of reliability and validity evidence for screening tools for cognitive and behavioral change in people with ALS? A systematic review. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, *25*(5-6), 437-451

69

69

Pharmacotherapy

- Disease-modifying therapies are intended to slow functional decline.
- Medications to manage symptoms.



70

70

Medication Side Effects

- Narcotics
  - Respiratory depression
- Diuretics
  - Dehydration, constipation
- Disease modifying medications
  - Taste changes
- Anticholinergics
  - Drowsiness, constipation

71

71

A-LS-PIKES Protocol

- Supports compassionate, clear communication for navigating complex and sensitive patient conversations.

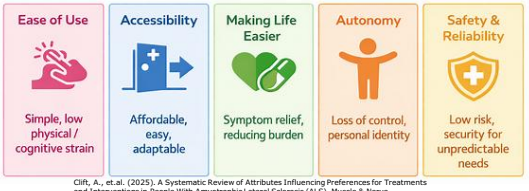
O'Connell, C., et.al. (2025). How to break the news in amyotrophic lateral sclerosis/motor neuron disease: practical guidelines from experts. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 26(1-2), 5-14.

72

72

Patient Preferences and End of Life

- Eating and Drinking with Acknowledged Risks (EDAR)
- Cognition may impact treatment decisions and compliance.
- Hospice care and dignity therapy



Clift, A., et.al. (2025). A Systematic Review of Attributes Influencing Preferences for Treatments and Interventions in People With Amyotrophic Lateral Sclerosis (ALS). Muscle & Nerve.

73

73

Conclusion

- While ALS remains a life-limiting disease, clinicians play a vital role in preserving dignity, prolonging function, and supporting meaningful participation for people living with ALS through compassionate, person-centered care.

74

74

1. Which of the following best describes Amyotrophic Lateral Sclerosis?
   A. Autoimmune demyelinating disorder of the central nervous system
   B. Progressive neurodegenerative disease of central and peripheral nervous system
   C. Chronic inflammatory condition of the peripheral nerves
   D. Reversible muscular disorder with episodic weakness

75

75

2. Approximately what percentage of the cases of ALS are familial/hereditary?
   A. 10%
   B. 30%
   C. 60%
   D. 90%

76

76

3. Which of the following environmental risk factors has been associated with the development of ALS?
   A. Smoking
   B. Microplastics
   C. Head injury
   D. Sedentary lifestyle

77

77

4. Which imaging modality measures water diffusion in the brain to identify white-matter microstructural damage?
   A. DTI
   B. Ultrasound
   C. EMG
   D. MRI

78

78

5. Early clinical signs of ALS typically include:
   A. Sensory loss and paresthesia
   B. Asymmetrical weakness and fasciculations
   C. Seizures and myoclonus
   D. Cognitive decline and apathy

79

79

6. Which domain is not assessed on the ALS Functional Rating Scale (ALSFRS-R)?
   A. Emotional lability
   B. Speech
   C. Fine motor tasks
   D. Respiratory function

80

80

7. What is the primary goal of respiratory training?
   A. Prevent aspiration during meals
   B. Improve oxygen saturation at rest
   C. Eliminate the need for non-invasive ventilation
   D. Delay the onset of ventilatory failure

81

81

8. Why is early screening for dysphagia critical in ALS management?
   A. Avoids overuse jaw injuries
   B. Ensures tongue strength for speech
   C. Minimizes risk of pneumonia due to aspiration
   D. Prevents TMJ dysfunction

82

82

9. Why is hypercaloric nutrition often recommended?
   A. Offset increased cholesterol production
   B. Support neural remyelination
   C. Reduce oxidative muscle metabolism
   D. Counteract muscle wasting and weight loss

83

83

10. The FAB can help identify what type of cognitive deficits?
   A. Visual memory
   B. Frontal executive function
   C. Language comprehension
   D. Procedural memory

84

84

Amyotrophic Lateral Sclerosis: Management Across the Disease Continuum

References

Aiello, E. N., Solca, F., Torre, S., Carelli, L., Ferrucci, R., Priori, A., ... & Poletti, B. (2023). Feasibility and diagnostics of the Frontal Assessment Battery (FAB) in amyotrophic lateral sclerosis. Neurological Sciences, 44(2), 587-592.

Al-Khayri, J. M., Ravindran, M., Banadka, A., Vandana, C. D., Priya, K., Nagella, P., & Kukkemane, K. (2024). Amyotrophic Lateral Sclerosis: Insights and New Prospects in Disease Pathophysiology, Biomarkers and Therapies. *Pharmaceuticals*, *17*(10), 1391.

Ansari, U., Wen, J., Karabala, M., Syed, B., Abed, I., Razick, D. I., & Lui, F. (2025). Analysis of Respiratory Muscle Strength Training in Amyotrophic Lateral Sclerosis (ALS) Patients: A Systematic Review. *Cureus*, *17*(2).

Benatar, M., Heiman-Patterson, T. D., Cooper-Knock, J., Brickman, D., Casaletto, K. B., Goutman, S. A., ... & Al-Chalabi, A. (2025). Guidance for clinical management of pathogenic variant carriers at elevated genetic risk for ALS/FTD. *Journal of Neurology, Neurosurgery & Psychiatry*, *96*(3), 209-218.

Benzo-Iglesias, M. J., Rocamora-Pérez, P., Valverde-Martínez, M. D. L. Á., García-Luengo, A. V., Benzo-Iglesias, P. M., & López-Liria, R. (2025). Efficacy of respiratory muscle training in improving pulmonary function and survival in patients with amyotrophic lateral sclerosis: a systematic review and meta-analysis. *Therapeutic advances in respiratory disease*, *19*, 17534666251346095.

Berry, J. D., Hagan, M., Zhang, J., Liu, Y., & Ciepielewska, M. (2025). Longer disease progression milestone-free time in people with amyotrophic lateral sclerosis treated versus not treated with intravenous edaravone: results from an administrative claims analysis. *Journal of Comparative Effectiveness Research*, *14*(2), e240007.

Bjelica, B., Bartels, M. B., Hesebeck-Brinckmann, J., & Petri, S. (2024). Non-motor symptoms in patients with amyotrophic lateral sclerosis: current state and future directions. *Journal of neurology*, *271*(7), 3953-3977.

Burke, K. M., Arulanandam, V., Scirocco, E., Royse, T., Hall, S., Weber, H., ... & Paganoni, S. (2025). Assistive Technology in ALS: A Scoping Review of Devices for Limb, Trunk, and Neck Weakness. *American Journal of Physical Medicine & Rehabilitation*, 10-1097.

Cheng, J., Niu, X., Li, H., Yang, Q., & Du, K. (2024). Evaluation of the therapeutic effects of rehabilitation therapy on patients with amyotrophic lateral sclerosis—a meta-analysis. *Frontiers in Neurology*, *15*, 1389146.

Clift, A., Rowen, D., Knox, L., Griffiths, A. W., & McDermott, C. J. (2025). A Systematic Review of Attributes Influencing Preferences for Treatments and Interventions in People With Amyotrophic Lateral Sclerosis (ALS). *Muscle & Nerve*.

Dave, K. D., Oskarsson, B., Yersak, J., Krauss, R., Heiman-Patterson, T., Lomen-Hoerth, C., ... & Babu, S. (2025). Contributions of neurologists to diagnostic timelines of ALS and thinkALS as an early referral instrument for clinicians. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, *26*(3-4), 215-224.

de Carvalho Vilar, M. D., Coutinho, K. M. D., de Lima Vale, S. H., Dourado Junior, M. E. T., de Medeiros, G. C. B. S., Piuvezam, G., ... & Leite-Lais, L. (2025). Evidence-Based Nutritional Recommendations for Maintaining or Restoring Nutritional Status in Patients with Amyotrophic Lateral Sclerosis: A Systematic Review. *Nutrients*, *17*(5), 782.

De Silva, U., Madanian, S., Olsen, S., Templeton, J. M., Poellabauer, C., Schneider, S. L., ... & Rubaiat, R. (2025). Clinical Decision Support Using Speech Signal Analysis: Systematic Scoping Review of Neurological Disorders. *Journal of Medical Internet Research*, *27*, e63004.

Dergai, O., Wuu, J., Koziczak-Holbro, M., Malaspina, A., Granit, V., Hernandez, J. P., ... & Benatar, M. (2025). Skeletal muscle biomarkers of amyotrophic lateral sclerosis: a large-scale, multi-cohort proteomic study. *medRxiv*, 2025-04.

Didcote, L., Vitoratou, S., Al-Chalabi, A., & Goldstein, L. H. (2024). What is the extent of reliability and validity evidence for screening tools for cognitive and behavioral change in people with ALS? A systematic review. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, *25*(5-6), 437-451.

Faller, K. M., Chaytow, H., & Gillingwater, T. H. (2025). Targeting common disease pathomechanisms to treat amyotrophic lateral sclerosis. *Nature Reviews Neurology*, 1-17.

Fenili, G., Scaricamazza, S., Ferri, A., Valle, C., & Paronetto, M. P. (2024). Physical exercise in amyotrophic lateral sclerosis: a potential co-adjuvant therapeutic option to counteract disease progression. *Frontiers in Cell and Developmental Biology*, *12*, 1421566.

Filippi, M., Ghirelli, A., Spinelli, E. G., & Agosta, F. (2025). A comprehensive update on neuroimaging endpoints in amyotrophic lateral sclerosis. *Expert Review of Neurotherapeutics*, *25*(4), 397-413.

Fouad, A. I., Ismail, A. K., Ali, A. N., Mansour, B. S., Salah Eldin, K., Ramadan, M. A., ... & Elsisy, M. (2025). Evolution of Wheelchair Technology: A Comprehensive Overview of History, Disabilities, Types and Control Mechanisms. *Advanced Sciences and Technology Journal*, *2*(2), 1-20.

García-Casanova, P. H., & Vázquez-Costa, J. F. (2025). Advances in the early diagnosis of amyotrophic lateral sclerosis. *Expert Review of Neurotherapeutics*, *25*(4), 415-425.

González-Sánchez, M., Ramírez-Expósito, M. J., & Martínez-Martos, J. M. (2025). Pathophysiology, Clinical Heterogeneity, and Therapeutic Advances in Amyotrophic Lateral Sclerosis: A Comprehensive Review of Molecular Mechanisms, Diagnostic Challenges, and Multidisciplinary Management Strategies. *Life*, *15*(4), 647.

Gratzer, A., Gdynia, N., Sasse, N., Beese, R., Winterholler, C., Bauer, Y., ... & Gdynia, H. J. (2025). Rehabilitation in Amyotrophic Lateral Sclerosis: Recommendations for Clinical Practice and Further Research. Journal of Clinical Medicine, 14(23), 8590.

Hamad, A. A., Alkhawaldeh, I. M., Nashwan, A. J., Meshref, M., & Imam, Y. (2025). Tofersen for SOD1 amyotrophic lateral sclerosis: a systematic review and meta-analysis. *Neurological Sciences*, 1-9.

Hamad, A. A., Amer, B. E., Abbas, N. B., Alnajjar, A. Z., & Meshref, M. (2024). Prevalence and correlates of fatigue in amyotrophic lateral sclerosis: a systematic review and meta-analysis. *Neurological Sciences*, *45*(2), 485-493.

Katerelos, A., Alexopoulos, P., Economou, P., Polychronopoulos, P., & Chroni, E. (2024). Cognitive function in amyotrophic lateral sclerosis: a cross-sectional and prospective pragmatic clinical study with review of the literature. *Neurological Sciences*, *45*(5), 2075-2085.

Katzeff, J. S., Bright, F., Phan, K., Kril, J. J., Ittner, L. M., Kassiou, M., ... & Kim, W. S. (2022). Biomarker discovery and development for frontotemporal dementia and amyotrophic lateral sclerosis. *Brain*, *145*(5), 1598-1609.

Kew, S. Y. N., Mok, S. Y., & Goh, C. H. (2024). Machine learning and brain-computer interface approaches in prognosis and individualized care strategies for individuals with amyotrophic lateral sclerosis: A systematic review. *MethodsX*, *13*, 102765.

Khan, A., Frazer-Green, L., Amin, R., Wolfe, L., Faulkner, G., Casey, K., ... & Gay, P. (2023). Respiratory management of patients with neuromuscular weakness: American college of chest physicians clinical practice guideline and expert panel report. *Chest*, *164*(2), 394-413.

Kleinerova, J., Chipika, R. H., Tan, E. L., Yunusova, Y., Marchand-Pauvert, V., Kassubek, J., ... & Bede, P. (2025). Sensory dysfunction in ALS and other motor neuron diseases: clinical relevance, histopathology, neurophysiology, and insights from neuroimaging. *Biomedicines*, *13*(3), 559.

Li, Z., & Kang, H. (2024). Efficacy of non-pharmacological interventions for individuals with amyotrophic lateral sclerosis: Systematic review and network meta-analysis of randomized control trials. *Scientific reports*, *14*(1), 11365.

Macpherson, C. E., Wani, D. K., Li, H., Rana, V., Blacutt, M., Bello-Haas, V. D., & Quinn, L. (2026). Physical Therapist Interventions for People With Amyotrophic Lateral Sclerosis Across Disease Stages: A Systematic Review of Efficacy. Physical Therapy, 106(1), pzaf142.

Magni, E., Hochsprung, A., Cáceres-Matos, R., Pabón-Carrasco, M., Heredia-Camacho, B., Solís-Marcos, I., & Luque-Moreno, C. (2024). Effects of Respiratory Training on Pulmonary Function, Cough, and Functional Independence in Patients with Amyotrophic Lateral Sclerosis. *Neurology International*, *16*(6), 1332-1342.

Maier, A., Kettemann, D., Weyen, U., Grehl, T., Schulte, P. C., Steinbach, R., ... & Spittel, S. (2025). Provision, cough efficacy and treatment satisfaction of mechanical insufflation-exsufflation in a large multicenter cohort of patients with amyotrophic lateral sclerosis. Scientific reports, 15(1), 7360.

McHenry, K. L. (2024). Airway clearance strategies and secretion management in amyotrophic lateral sclerosis. *Respiratory Care*, *69*(2), 227-237.

Myrou, A., Barmpagiannos, K., Ioakimidou, A., & Savopoulos, C. (2025). Molecular Biomarkers in Neurological Diseases: Advances in Diagnosis and Prognosis. *International Journal of Molecular Sciences*, *26*(5), 2231.

National Academies of Sciences, Engineering, and Medicine. 2024. Living with ALS. Washington, DC: The National Academies Press. https://doi.org/10.17226/27739

Newell, M. E., Aravindan, A., Babbrah, A., & Halden, R. U. (2025). Epigenetic Biomarkers Driven by Environmental Toxins Associated with Alzheimer's Disease, Parkinson's Disease, and Amyotrophic Lateral Sclerosis in the United States: A Systematic Review. *Toxics*, *13*(2), 114.

O'Connell, C., Kavanaugh, M. S., Cummings, C., & Genge, A. (2025). How to break the news in amyotrophic lateral sclerosis/motor neuron disease: practical guidelines from experts. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, *26*(1-2), 5-14.

Oh, J., An, J., Park, K., & Park, Y. (2024). Psychosocial interventions for people with amyotrophic lateral sclerosis and motor neuron disease and their caregivers: a scoping review. *BMC nursing*, *23*(1), 75.

Orlova, A., Malygin, Y., Gofman, A., Sotulenko, S., Gandalian, V., Kartashov, I., ... & Jakovljevic, V. (2024). Survival Prognostic Factors of Non-Invasive Ventilation in Amyotrophic Lateral Sclerosis: A Systematic Review. *Life*, *14*(12), 1664.

Pasqualucci, E., Angeletti, D., Rosso, P., Fico, E., Zoccali, F., Tirassa, P., ... & Severini, C. (2025). Management of dysarthria in amyotrophic lateral sclerosis. *Cells*, *14*(14), 1048.

Qin, J., Wang, X., Fan, G., Zhang, W., Wu, X., Wang, B., & Liu, Y. (2024). Identifying amyotrophic lateral sclerosis using diffusion tensor imaging, and correlation with neurofilament markers. *Scientific reports*, *14*(1), 28110.

Raymond, J., Howard, I. M., Berry, J., Larson, T., Horton, D. K., & Mehta, P. (2025). Head Injury and Amyotrophic Lateral Sclerosis: Population-Based Study from the National ALS Registry. *Brain Sciences*, *15*(2), 143.

Ren, S., Che, X., Hu, S., Feng, X., Zhang, J., & Shi, P. (2025). The effect of exercise intervention on amyotrophic lateral sclerosis: A systematic review and meta-analysis. *Frontiers in Neurology*, *16*, 1499407.

Rojas-López, J. C., Estrada-Gualdron, P. I., Ramírez-Guerrero, S., Velásquez-Cárdenas, M. J., Redondo-Escobar, J., Vargas-Arenas, S., ... & Palacios-Espinosa, X. (2024). Efficacy of pain

management strategies in adults with Amyotrophic Lateral Sclerosis (ALS): A Systematic Review. *Neurological Sciences*, *45*(12), 5591-5604.

Rosca, C. M., & Stancu, A. (2025). A Review of Neuro-ML Breakthroughs in Addressing Neurological Disorders. *Applied Sciences*, *15*(10), 5442.

Silva, S. T., Costa, I. M., Souza, A. A., Pondofe, K., Melo, L. P., Resqueti, V. R., ... & Ribeiro, T. S. (2024). Physical therapy for the management of global function, fatigue and quality of life in amyotrophic lateral sclerosis: systematic review and meta-analyses. *BMJ open*, *14*(8), e076541.

Souza, A. A. D., Silva, S. T. D., Régis, A. M. P., Aires, D. N., Pondofe, K. D. M., Melo, L. P. D., ... & Ribeiro, T. S. (2025). Muscle strengthening in individuals with Amyotrophic Lateral Sclerosis: a systematic review with meta-analyses. *PloS one*, *20*(4), e0320788.

Tan, H. H., Nitert, A. D., van Veenhuijzen, K., Dukic, S., van Zandvoort, M. J., Hendrikse, J., ... & van den Berg, L. H. (2025). Neuroimaging correlates of domain-specific cognitive deficits in amyotrophic lateral sclerosis. NeuroImage: Clinical, 103749.

Urushitani, Makoto, Hitoshi Warita, Naoki Atsuta, Yuishin Izumi, Osamu Kano, Toshio Shimizu, Yuki Nakayama et al. "The clinical practice guideline for the management of amyotrophic lateral sclerosis in Japan—update 2023." *Rinsho Shinkeigaku* 64, no. 4 (2024): 252-271.

Van Damme, P., Al-Chalabi, A., Andersen, P. M., Chiò, A., Couratier, P., De Carvalho, M., ... & Weber, M. (2024). European Academy of Neurology (EAN) guideline on the management of amyotrophic lateral sclerosis in collaboration with European Reference Network for Neuromuscular Diseases (ERN EURO-NMD). *European journal of neurology*, *31*(6), e16264.

Vidovic, M., Lapp, H. S., Weber, C., Plitzko, L., Seifert, M., Steinacker, P., ... & Günther, R. (2024). Comparative analysis of neurofilaments and biomarkers of muscular damage in amyotrophic lateral sclerosis. *Brain communications*, *6*(5), fcae288.

Wąsik, J., Kurkiewicz, W., Woźniak, O., Mikulska, J., Gryboś, K., Lewandowski, M., ... & Kwiatkowska, Z. (2025). The role of diet and nutritional deficiences in neurodegenerative diseases. *Quality in Sport*, *41*, 60334-60334.

Zhong, R., Rua, M. T., & Wei-LaPierre, L. (2024). Targeting mitochondrial Ca2+ uptake for the treatment of amyotrophic lateral sclerosis. *The Journal of Physiology*, *602*(8), 1519-1549.