NEUROLOGICAL SYSTEM · DEGENERATIVE DISORDER

Amyotrophic Lateral Sclerosis *(ALS)*

Also known as *Lou Gehrig's disease*. A progressive, fatal neurodegenerative disorder destroying upper and lower motor neurons — cognition, sensation, and sphincter control remain intact. Respiratory failure is the leading cause of death.

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i **Content sourcing note:** ALS was not included in the uploaded reference notes for this library. Content on this card is drawn from standard NGN NCLEX 2026 references — *Saunders Comprehensive Review*, *ATI Adult Medical-Surgical Nursing*, *Lippincott Manual of Nursing Practice*, and *ALS Association clinical guidelines (2024–2025)* — and follows the established design pattern of the Visual Study Library.

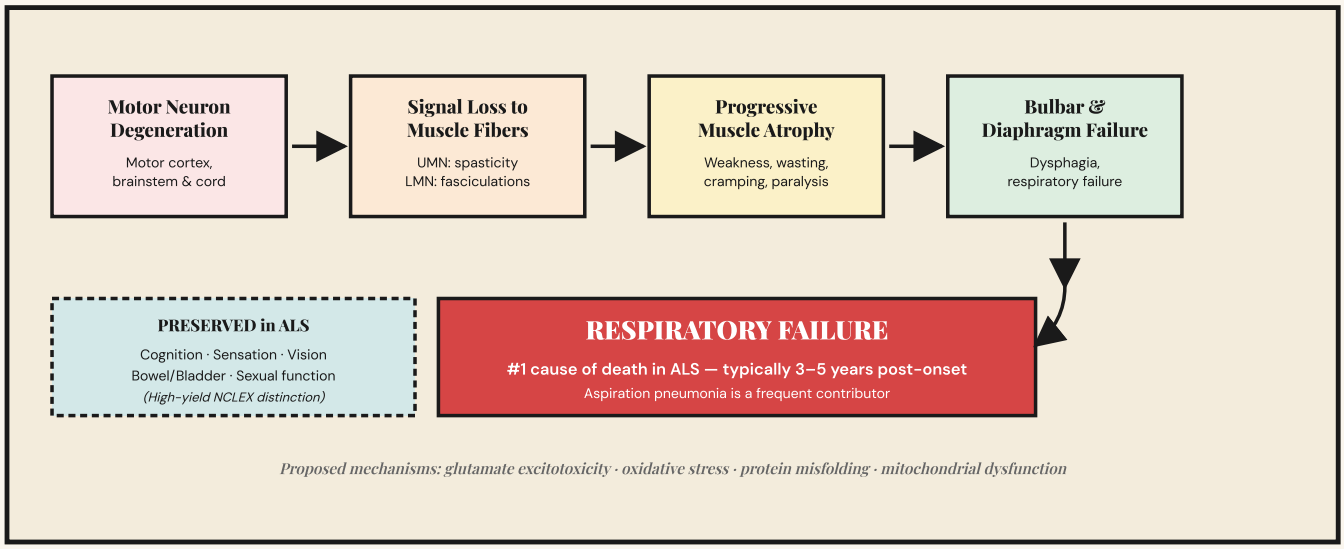
1 Definition & Overview

- **What it is:** A progressive degeneration of **upper motor neurons** (motor cortex) and **lower motor neurons** (brainstem, spinal cord) leading to muscle weakness, atrophy, paralysis, and death.
- **Onset:** Typically ages 40–70; slightly more common in males. Most cases are sporadic; ~5–10% are familial (SOD1 gene).
- **Prognosis:** Median survival **3–5 years from symptom onset**. There is **no cure** — treatment slows progression and supports quality of life.
- **What is PRESERVED:** Cognition (in most), sensation, vision/hearing, bowel/bladder, and sexual function. This distinguishes ALS from many other neurodegenerative diseases.

⚠ **Tested fact: ALS is purely a *motor* disease. If an NCLEX question describes loss of sensation, dementia, or incontinence as the primary feature → it is NOT ALS.**

2 Pathophysiology (Simplified Flow)

Motor neurons in the brain and spinal cord progressively die. With no signal reaching the muscle, the muscle wastes away. Both **upper** (UMN) and **lower** (LMN) motor neurons are affected — producing a unique mix of *spasticity* (UMN) and *flaccid weakness with fasciculations* (LMN).



3 Signs & Symptoms

EARLY Onset Signs

- **Limb weakness** — often asymmetric, starts in hands (dropping things, buttoning) or feet (foot drop, tripping)
- **Fasciculations** — visible muscle twitching (classic LMN sign)
- **Muscle cramping** and stiffness
- **Dysarthria** — slurred, nasal speech
- **Mild dysphagia** — trouble with thin liquids first
- **Fatigue** with normal activity

LATE & Advanced Signs

- **Progressive paralysis** — flaccid + spastic mix
- **Severe dysphagia** → aspiration risk
- **Sialorrhea** — excessive drooling (can't manage saliva)
- **Anarthria** — total loss of speech
- **Dyspnea, orthopnea, weak cough** → respiratory failure
- **Pseudobulbar affect** — uncontrolled laughing/crying
- **Complete dependence** for ADLs; *cognition still intact*

RED FLAGS — Report immediately: Declining O_2 sat, weak/ineffective cough, choking during meals, accessory muscle use, paradoxical breathing, drop in FVC (forced vital capacity) below 50%, morning headaches (CO_2 retention).

PRESERVED — Do NOT expect these losses

Cognition (most patients) · Sensation (touch, pain, temperature) · Eye movement (often spared until very late) · Bowel and bladder control · Sexual function. **This pattern is the NCLEX fingerprint of ALS.**

4 Priority Nursing Interventions (ABC Framework)

A — Airway

- **Suction** oral secretions PRN (yankauer)
- Keep **HOB ≥ 30–45°**
- Encourage **chin-tuck** while swallowing
- Monitor for choking/aspiration with every meal

B — Breathing

- **Monitor FVC, NIF, SpO_2, ABGs**
- Provide **BiPAP / NIV** as ordered (slows decline)
- Teach **incentive spirometry** & cough-assist
- Prepare for **tracheostomy** discussion early
- Pneumococcal & influenza vaccines

C — Circulation & Care

- **DVT prophylaxis** (SCDs, mobility, anticoag)
- Reposition q2h to **prevent skin breakdown**
- Passive ROM to prevent contractures
- Monitor weight & intake closely

NUTRITION — Aspiration Precautions

- Sit **upright 90°** for meals & 30 min after
- **Thickened liquids** (honey/nectar consistency)
- Soft, moist foods; small bites; chin-tuck
- Suction at bedside ready at all times
- **PEG tube** placed *early* — before FVC < 50% (safer placement)

COMMUNICATION & PSYCHOSOCIAL

- Provide **communication boards**, eye-gaze devices, speech apps
- Allow **extra time**; never finish patient's sentences
- Address **depression, anxiety, grief** — refer to support
- Initiate **advance directives** & palliative consult *early*
- Caregiver education and respite resources

5 Pharmacology — Disease-Modifying & Symptom Drugs

Drug / Class	Mechanism (simple)	Key Side Effects	Nursing Considerations
Riluzole ***(Rilutek)*** **Glutamate inhibitor**	Blocks excess glutamate → reduces motor neuron damage. **First-line disease-modifier.**	Hepatotoxicity, dizziness, nausea, weakness, neutropenia	Monitor **LFTs monthly × 3, then q3 mo**. Take on **empty stomach** (1 hr before / 2 hr after meals). Avoid alcohol. Extends survival ~3 months.
Edaravone ***(Radicava)*** **Antioxidant / free-radical scavenger**	Reduces oxidative stress on motor neurons → slows functional decline.	Bruising, gait disturbance, headache, sulfite allergy reactions	IV infusion (60 min) or oral suspension. Cyclic dosing schedule. Check for **sulfite allergy** before first dose.
Sodium phenylbutyrate / Taurursodiol ***(Relyvrio)***	Reduces neuronal cell death (mitochondrial & ER stress pathways).	Diarrhea, abdominal pain, nausea	Note: withdrawn from US market in 2024 after Phase 3 failure — may still appear in older NCLEX banks. *Verify current availability.*
Baclofen / Tizanidine **Muscle relaxants**	Reduces UMN spasticity.	Sedation, weakness, hypotension	Avoid abrupt discontinuation of baclofen (withdrawal seizures). Fall precautions.
Glycopyrrolate / Atropine drops / Botulinum toxin	Anticholinergic — reduces saliva production for sialorrhea.	Dry mouth, constipation, urinary retention, confusion	Watch for thick secretions (paradoxically worsens airway). Provide oral care.
Dextromethorphan / Quinidine ***(Nuedexta)***	Treats **pseudobulbar affect** (uncontrolled laughing/crying).	Dizziness, diarrhea, QT prolongation	Monitor ECG for QT prolongation. Avoid with other QT-prolonging drugs.
Opioids / Benzodiazepines (end-stage)	Relieves dyspnea, anxiety, pain in palliative phase.	Respiratory depression, sedation, constipation	Comfort-focused care; titrate to symptom relief, not numbers. Bowel regimen.

6 NCLEX Test Traps ⚠

TRAP Confusing ALS with **Alzheimer's, MS, or Parkinson's** because they're all neuro.	→	CORRECT ALS = **motor only**. Cognition, sensation, bowel/bladder are **intact**. If the stem describes memory loss or numbness — it's NOT ALS.
TRAP Picking "**turn and reposition**" or "**provide emotional support**" as the priority intervention.	→	CORRECT **Airway and aspiration risk always win.** Suction, HOB up, swallow precautions, BiPAP are higher priority than skin or psychosocial care.
TRAP Assuming the patient **can't understand** because they can't speak.	→	CORRECT Cognition is **preserved**. Speak directly to the patient, allow processing time, use communication devices. *Never* talk over them.
TRAP Waiting until severe weight loss or respiratory failure to discuss a **feeding tube**.	→	CORRECT Insert **PEG tube EARLY** — before FVC drops below 50%. Late placement carries far higher procedural risk.
TRAP Choosing "**encourage independence with ADLs**" when the patient is exhausted.	→	CORRECT **Energy conservation** is the principle. Assist with ADLs to preserve strength for breathing and eating. Pushing independence worsens fatigue.
TRAP Selecting "**full code, aggressive treatment**" as the default goal of care.	→	CORRECT Initiate **advance directives and palliative care discussions early** — at diagnosis. The patient is cognitively intact and has the right to make decisions about ventilation, tube feeding, and DNR.

7 Patient & Family Education

Daily Living & Safety

- **Energy conservation:** rest before activity, plan high-priority tasks for morning
- **Aspiration precautions:** upright meals, thickened liquids, small bites, chin-tuck
- **Home modifications:** ramps, grab bars, hospital bed, hoyer lift, suction setup
- Use of **AFO braces** for foot drop; walker → wheelchair → power chair
- Skin checks; turn schedule; pressure-relieving cushions

Planning Ahead

- **Advance directive** & living will *while cognition and speech are intact*
- Discuss **BiPAP, tracheostomy, PEG tube, DNR** early
- Connect with **ALS Association**, hospice/palliative services, support groups
- Communication backup plan (eye-gaze, letter board, speech-banking apps)
- **Caregiver education:** suctioning, tube feeds, transfers, signs of distress
- Vaccinations: **flu yearly, pneumococcal, COVID** boosters

8 Memory Boosters & Mnemonics

A·L·S

- **A** Atrophy of muscles (motor only)
- **L** Loss of motor neurons (UMN + LMN)
- **S** Sparing of senses, cognition, bowel/bladder

BREATHE

- **B** BiPAP / NIV early
- **R** Riluzole + LFT monitoring
- **E** Energy conservation
- **A** Aspiration precautions
- **T** Tube feeds (PEG) early
- **H** HOB up, suction ready
- **E** End-of-life planning

Pattern Recognition Tips

- ☐ **UMN signs** → *spasticity, hyperreflexia, Babinski +*
- ☐ **LMN signs** → *flaccidity, atrophy, fasciculations, hyporeflexia*
- ☐ ALS uniquely has **BOTH** at the same time in the same patient.
- ☐ "Mind trapped in a failing body" = classic ALS description
- ☐ "Lou Gehrig's disease" = trigger word on NCLEX

Quick Drug Recall

- ☐ **Ri**luzole = **Ri**verse-the-glutamate (slows progression)
- ☐ **Edara**vone = **era**se oxidative stress
- ☐ **Baclofen** = **B**locks **B**ad spasticity (no abrupt stop!)
- ☐ **Glycopyrrolate** = "Goodbye-spit" (sialorrhea control)

9 NCJMM 6-Step Clinical Judgment Scenario

SCENARIO: A 58-year-old male with ALS (diagnosed 2 years ago) presents to clinic with his wife. He uses a walker and a communication tablet because his speech is slurred. His wife reports he has been coughing during meals over the past week, lost 6 lb in 2 weeks, and "wakes up with terrible headaches." His SpO_2 is 91% on room air, FVC is 48% of predicted, RR 24, lungs with bilateral basilar crackles, T 38.1°C (100.6°F). He is alert and oriented and pulls out his tablet to type: *"I'm scared to choke."*

1 Recognize Cues

Relevant cues: Coughing with meals · 6 lb weight loss · Morning headache (suggests *CO_2 retention*) · SpO_2 91% · **FVC 48%** (below threshold) · Tachypnea · Basilar crackles · Low-grade fever · Patient anxiety about choking. **Cognition is intact.**

2 Analyze Cues

The cluster points to **two converging problems:** (1) **aspiration** — coughing with meals + weight loss + fever + crackles = likely aspiration pneumonia; (2) **impending respiratory failure** — FVC < 50%, morning headache, low SpO_2. These problems amplify each other and threaten airway. Anxiety is real and rational.

3 Prioritize Hypotheses

#1 Highest priority: Respiratory compromise with possible aspiration pneumonia (ABC — airway/breathing). **#2:** Nutritional decline / dysphagia. **#3:** Psychosocial distress & advance care planning. Lower-priority concerns: skin integrity, ADL assistance.

4 Generate Solutions

Expected interventions: **elevate HOB 45–90°**, supplemental O_2 titrated to SpO_2 > 92%, obtain ABG & chest X-ray, suction setup at bedside, NPO until swallow eval, start **BiPAP** per protocol, consult speech-language pathology, **discuss PEG tube placement** (FVC is borderline for safe insertion), antibiotics if pneumonia confirmed, reinforce advance directive review, palliative care referral.

5 Take Action

Nurse immediately raises HOB, applies O_2 via nasal cannula at 2 L/min and reassesses SpO_2, places patient NPO, sets up suction, notifies HCP of FVC drop and clinical findings, draws ordered labs/ABG, prepares for BiPAP trial, contacts SLP and dietitian, uses the communication tablet to acknowledge the patient's fear and explain each action, and involves wife in planning.

6 Evaluate Outcomes

Expected positive outcomes: SpO_2 > 92%, RR returns to 12–20, ABG shows improved or stable CO_2, patient reports decreased dyspnea/anxiety, no further choking episodes, weight stabilizes after PEG initiation, advance directive updated and on chart. **Reassess in 15–30 min**; escalate to higher level of care if respiratory status worsens or BiPAP not tolerated.