Acute and Chronic Neurological Diseases: Quick Notes

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Functional Health Patterns (53%): This domain involves applying the nursing process to optimize the restoration and preservation of the patient's health and holistic well-being across the lifespan. It also includes promoting optimal psychosocial patterns and coping and stress management skills of the patients and caregivers, optimizing the patient's functional ability, managing the patient's neurological and other complex medical conditions, promoting optimal nutrition and hydration, optimizing the patient's elimination patterns, and optimizing the patient's sleep and rest patterns.

Neurological Conditions

- Multiple Sclerosis
- Parkinson's Disease
- Guillain-Barré Syndrome
- Myasthenia Gravis
- Amyotrophic Lateral Sclerosis
- Postpolio Syndrome

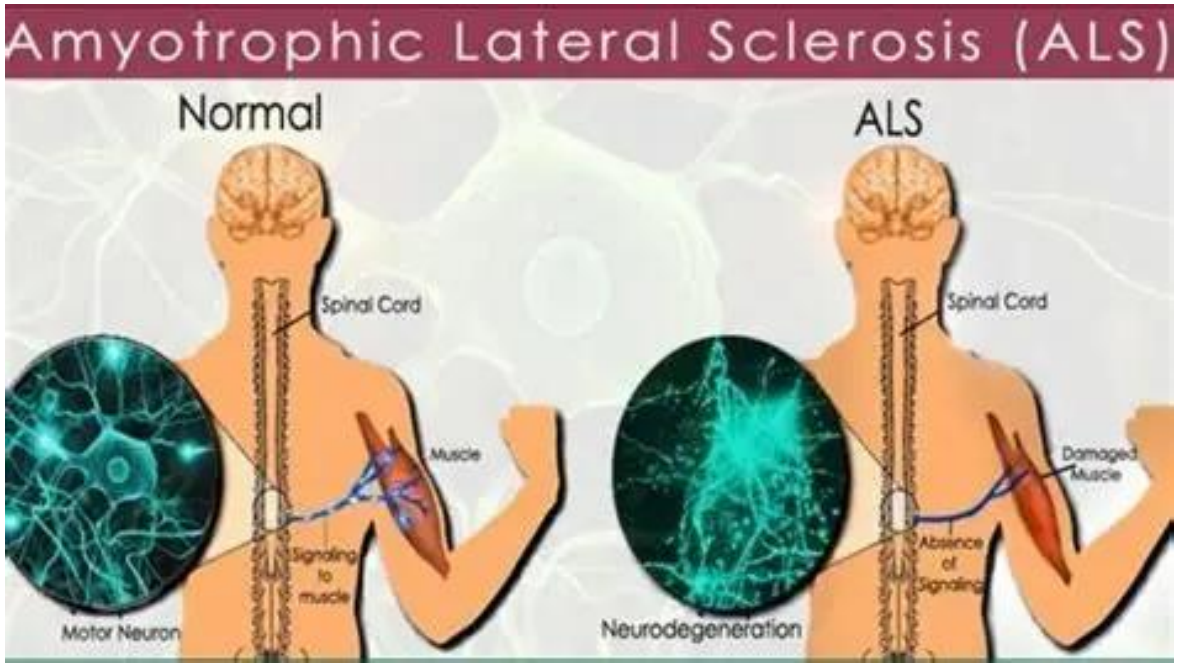
Objectives

Participants will be able to:

- Describe scope of each neurological disease/condition
- State Symptoms of each neurological disease/condition
- Identify the Goals of the Rehabilitation Nurse related the specific neurological disease/condition
- Describe the Nursing Interventions for each neurological condition

Amyotrophic Lateral Sclerosis

- Lou Gehrig's Disease



Amyotrophic Lateral Sclerosis (ALS)

Category	Key Points
Global Prevalence	~5 per 100,000
Incidence	~1.5–2.5 per 100,000/year
Gender Distribution	More common in **men** (1.2–1.5:1), but gap narrows with age
Age of Onset	Typically between **55–75 years**
Ethnicity & Race	Higher in **White populations**; lower in Black and Asian populations
Geographic Patterns	Higher in Europe, North America
Trends	Slight increase in incidence; survival improving with supportive care

Common Symptoms: Amyotrophic Lateral Sclerosis

- Progressive muscle weakness
- Fasciculations (muscle twitching)
- Spasticity and stiffness
- Dysarthria (slurred speech)
- Dysphagia (difficulty swallowing)
- Respiratory difficulty
- Cognitive changes (in some cases)

Management / Treatment: ALS

🧪 **Disease-Modifying**

- **Riluzole** – prolongs survival
- **Edaravone** – slows functional decline

🩺 **Supportive Care/ Symptom management**

- Non-invasive ventilation (BiPAP)
- PEG feeding for dysphagia
- Multidisciplinary care (PT, OT, speech, palliative)

🧠 **Emerging**

- Ongoing trials in gene therapy, stem cells, and neuroprotective agents

Amyotrophic Lateral Sclerosis (ALS)

🩺 **Rehabilitation Nursing Focus:**

- **Assistive technology**: Communication devices, mobility aids.
- **Respiratory care**: Non-invasive ventilation, secretion management.
- **Nutritional support**: PEG feeding when dysphagia progresses.
- **Palliative care**: Symptom relief, advance care planning.
- **Psychosocial support**: For patient and caregivers coping with progressive decline.

Amyotrophic Lateral Sclerosis

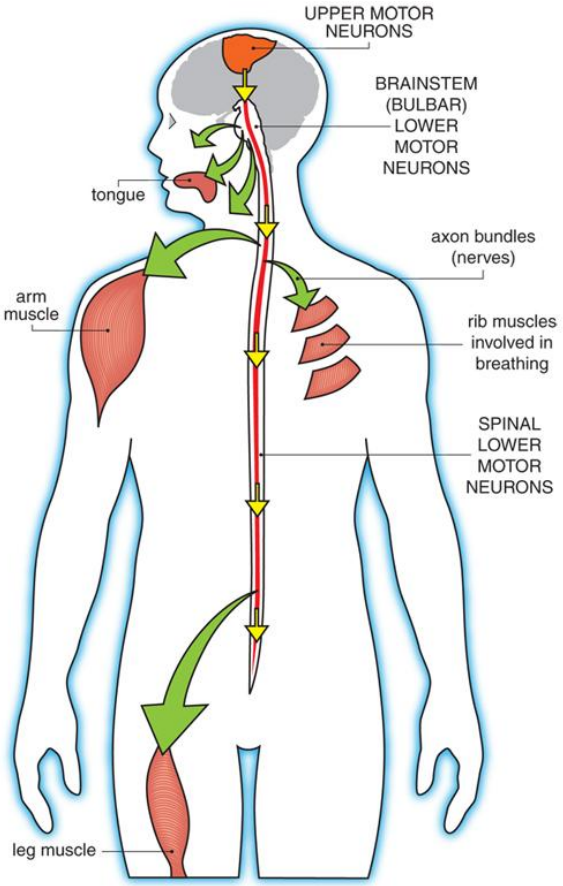
- Neurological disorder with progressive degeneration of skeletal muscle motor/nerve cells throughout the nervous system
- No cerebellar effects
- No sensory loss- posterior nerve roots not affected
- Anterior horn cells of S2 not affected, rare bowel & bladder deficits
- Movement disorder is profound

Variations in Disease Progression

- **Functional Loss may start with Upper-motor neurons, lower-motor neurons, bulbar symptoms only or a combination**
- **Onset is often subtle- first symptoms maybe disregarded**
- **No Single Diagnostic Test to diagnosis ALS**
- **No know Prevention and No known Cure**
- **Treatments to assist in slowing deterioration**

Amyotrophic Lateral Sclerosis

- Symptoms-
  - Upper motor neuron disease
    - Weakness
    - spasticity
    - hyperactive reflexes
    - hypertonicity
    - Disuse atrophy
  - Lower motor neuron disease
    - Weakness
    - Flaccidity
    - Hypoactive reflexes
    - Hypotonicity and muscle atrophy



ALS

- Intellectual ability, vision, hearing, and sensation are not generally affected
- Some people do experience cognitive deficits
  - Cognitive impairment can be
    - Frontotemporal dementia with cognitive decline and increased apathy
    - Mild cognitive impairments with no detectable progression

Amyotrophic Lateral Sclerosis

What is ALS?

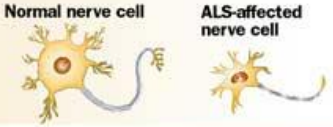
ALS (Amyotrophic Lateral Sclerosis), also known as Lou Gehrig's disease, is a fatal disease of the nervous system, characterized by progressive muscle weakness resulting in paralysis.

What are motor neurons?

Motor neurons are nerve cells in the brain and spinal cord that attach to muscles and control voluntary movement.

How does ALS progress?

When motor neurons gradually degenerate and die, the muscles no longer receive nerve impulses. As a result of the nerve death, the muscles shrink and waste away.



A closer look at a healthy nervous system

Nervous system

The basic unit of the nervous system is a highly specialized cell, known as a neuron. Its main purpose is to transport messages from one part of the body to another in the form of nerve impulses.

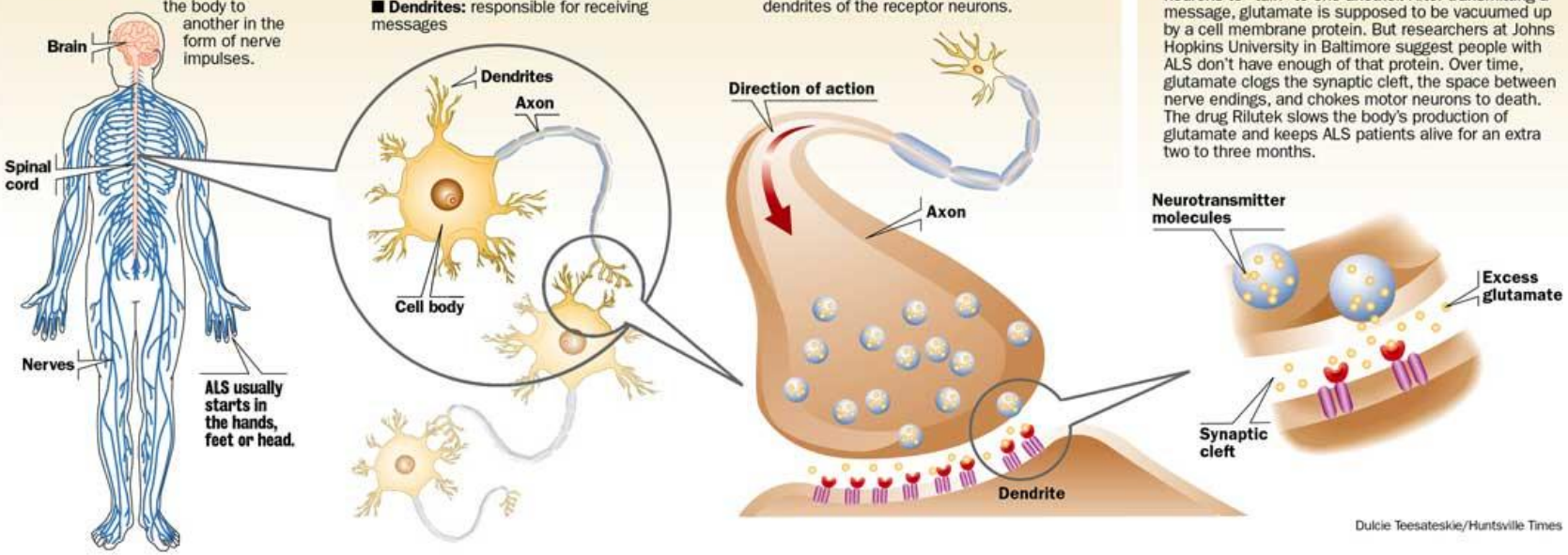
Motor neuron

A motor neuron is made up of three main functional parts.

- **Cell body:** biosynthetic center of the cell
- **Axon:** responsible for sending messages
- **Dendrites:** responsible for receiving messages

Nerve impulse

A nerve impulse is transmitted when the terminal fibers of one neuron's axon release chemicals called neurotransmitters that attach to dendrites of the receptor neurons.



A possible cause of ALS: Too much glutamate

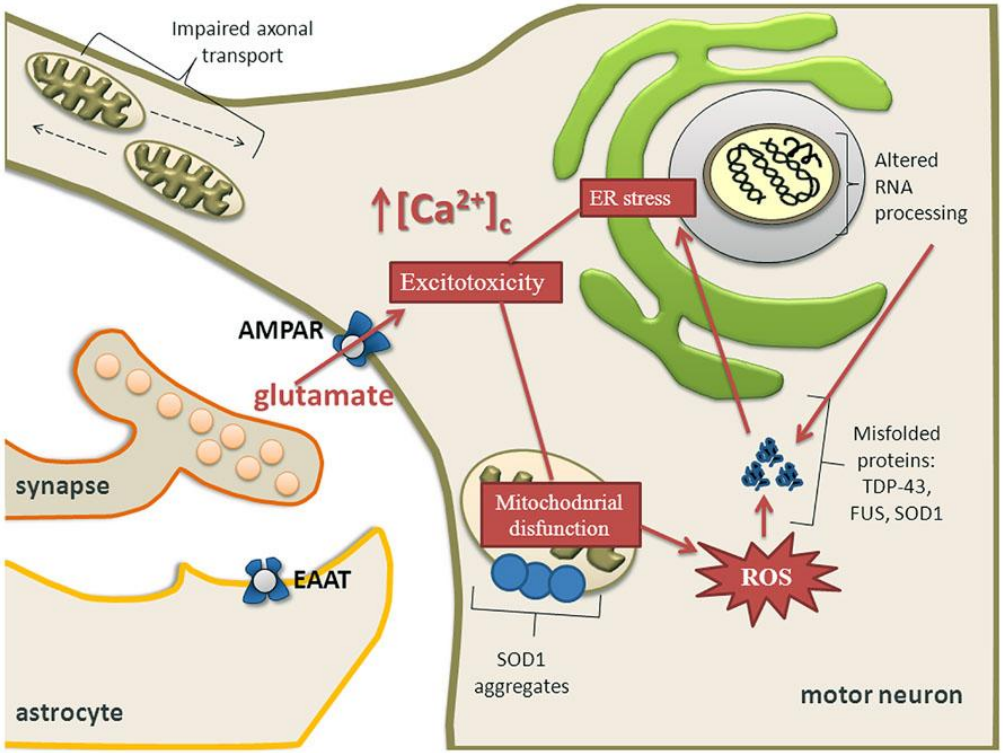
Scientists aren't sure what causes ALS, but glutamate poisoning is a popular theory. Glutamate is an amino acid that acts as a neurotransmitter, allowing motor neurons to "talk" to one another. After transmitting a message, glutamate is supposed to be vacuumed up by a cell membrane protein. But researchers at Johns Hopkins University in Baltimore suggest people with ALS don't have enough of that protein. Over time, glutamate clogs the synaptic cleft, the space between nerve endings, and chokes motor neurons to death. The drug Rilutek slows the body's production of glutamate and keeps ALS patients alive for an extra two to three months.

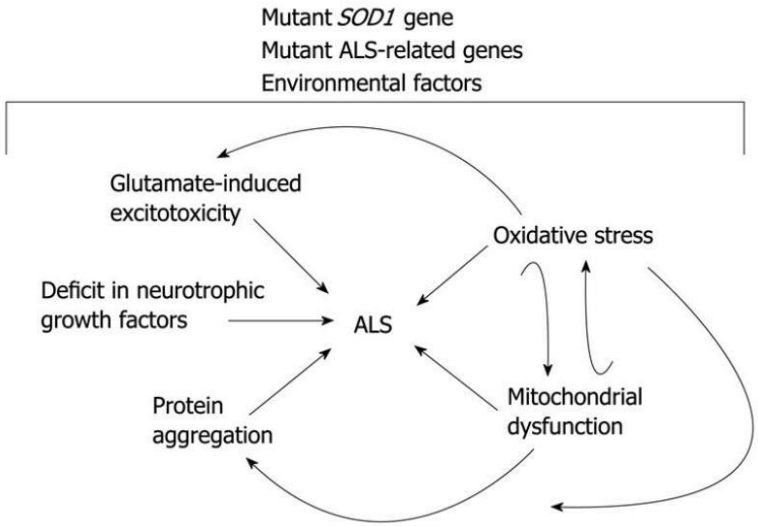
Dulcie Teesateskie/Huntsville Times

Etiology

- Sporadic , unkown cause
- Genetic in 5-10% of cases-

an inherited autosomal trait





Etiology

- Familial ALS: Approximately 5-10% of ALS cases are familial, meaning they have a genetic basis. Mutations in several genes, such as C9orf72 and SOD1, have been identified as contributing to familial ALS. These mutations can lead to the degeneration of motor neurons, which are crucial for muscle control.
- Sporadic ALS: The majority of ALS cases (about 90-95%) are sporadic, occurring randomly without a clear family history; researchers are investigating potential genetic predispositions that may increase susceptibility to the disease.
- Toxin Exposure: Some studies suggest that exposure to environmental toxins, such as heavy metals (e.g., lead), pesticides, and other chemicals, may be linked to an increased risk of developing ALS. However, no specific agent has been consistently associated with the disease.
- Lifestyle Factors: Smoking has been identified as a potential risk factor, particularly among women after menopause. Other Considerations
- Age and Sex: The risk of developing ALS increases with age, particularly between the ages of 60 and 80. Men are more likely to develop ALS than women before age 70, but this difference diminishes in older age groups.
- Military Service: Some studies indicate that military veterans may have a higher risk of ALS, potentially due to exposure to environmental toxins during service.

Rehabilitation Goals

- Prevent complications of immobility
- Strengthen unaffected muscles
- Saliva management
- Dysphasia
- Aspiration prevention, Assisted Cough
- Communication, plan early before need for ventilator
- Adaptive techniques and equipment
- Depression, coping with progressive loss
- Preparation for disease progression
- Preparation for death

Focus on Palliative Care

- Key Decisions must be discussed in advance of crisis
  - G-Tube
  - Mechanical Ventilation
  - Advance Directives
- Teaching Plan for Patient and Caregiver/Family
  - Symptom Management
  - Reduce Complications
- Assistive Devices to maintain independence
- Referrals for support groups and other resources

Nursing Process

- Assessment
  - Full Health History
  - Family Incidence of ALS
  - Onset of Symptoms and Progression
- Current Function & ADLs
  - Gait, strength and stability
  - Flaccidity, spasticity
  - Eval Swallowing and chewing
  - Eval Respiratory Status
  - Bowel and Bladder function
  - Skin Assessment
  - Observe Family Interactions

Plan of Care

- **Nursing Diagnoses**
  - Impaired physical mobility re: muscle wasting, weakness and spasticity
  - Self Care deficit
  - Impaired Communication re: impairment of muscles for speech
  - Ineffective breathing pattern re: impairment of diaphragm and accessory muscles

Plan of Care

- **Nursing Diagnoses**
  - Altered nutrition: Less than required re: bulbar muscles
  - Potential for anxiety
  - Risk for ineffective coping
  - Interrupted family processes re: change in health of family member, modifications of family roles and foreseen loss of family member
  - Risk for caregiver strain

Interventions

- Maintain Independence
- Limit complications from progressive loss of function
  - Ineffective breathing
  - Impaired swallowing and decrease in nutritional intake
  - Impaired communication
  - Maintain skin integrity
  - Patient and Family Education and support in decision making
  - Pain management

New Treatment Guidelines 2023 for ALS

- The main recommendations concerning disease-modifying therapies are the following:

  Riluzole should be offered as a lifelong treatment option to all ALS patients at diagnosis. "A single daily dose of 50 mg can already be effective," added Prof. Van Damme.

- Cell-based therapies are not recommended outside the context of clinical trials until positive phase 3 trial data is available.
- Edaravone and AMX0035 both receive temporary recommendations; the guideline committee wants to await the phase 3 trial results before issuing a final recommendation.
- Tofersen should be offered as first-line treatment in patients with progressive ALS caused by mutations in the superoxide dismutase 1 (*SOD1*) gene. The possibility of serious adverse events should be discussed with the patient.

Treatment for ALS

- Medications can be prescribed to help manage symptoms of ALS:
  - Muscle cramps
  - Stiffness
  - Excess saliva and phlegm
  - Pseudobulbar affect (involuntary or uncontrollable episodes of crying and/or laughing, or other emotional displays).

- Drugs also are available to help individuals with:
  - Pain
  - Depression
  - Sleep disturbances
  - Constipation

Non-Pharmacological Treatment

- **Communications support**
  - As ALS progresses, speech therapists can help people maintain the ability to communicate.
  - Devices such as **computer-based speech synthesizers** use eye-tracking technology and can help people develop ways for responding to yes-or-no questions with their eyes or by other nonverbal means
  - A **brain-computer interface (BCI)** is a system that allows individuals with ALS to communicate or control equipment such as a wheelchair using only brain activity.
- **Nutritional support**
  - Small meals that provide enough calories, fiber, and fluid
  - Avoid foods that are difficult to swallow
  - Suction devices
  - Feeding tube
- **Breathing support**
  - Noninvasive ventilation (NIV) refers to breathing
  - Mechanical ventilation (respirators)
- **Rehabilitation Team**
  - Physical Therapy
  - Occupational therapy
  - Speech Therapy