

# Acute and Chronic Neurological Diseases: Quick Notes



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# Neurological Conditions

## ▶ Amyotrophic Lateral Sclerosis

# 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.

Normal nerve cell



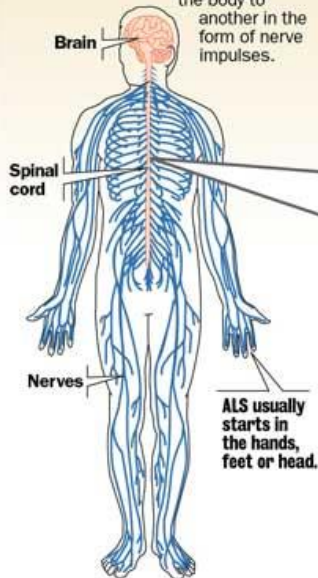
ALS-affected nerve cell



## A closer look at a healthy nervous system

### Nervous system

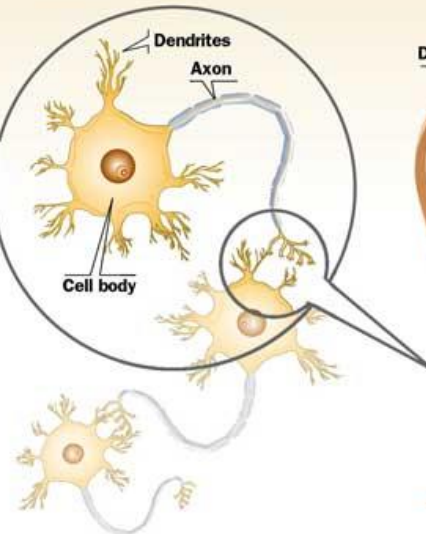
The basic unit of the nervous system is a highly specialized cell, known as a neuron. Its main purpose is to transport messages from one part of the body to another in the form of nerve impulses.



### Motor neuron

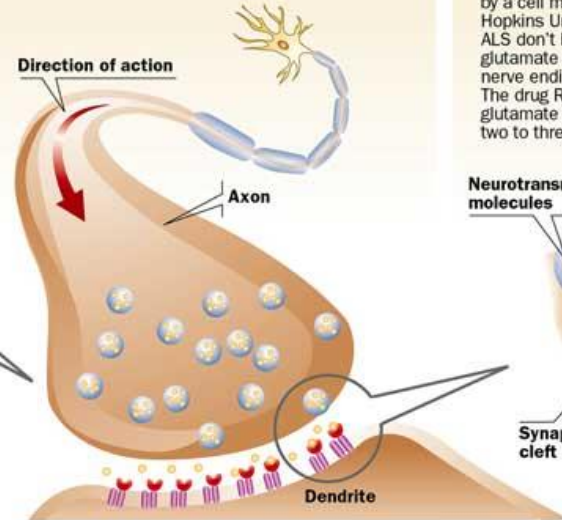
A motor neuron is made up of three main functional parts.

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



### Nerve impulse

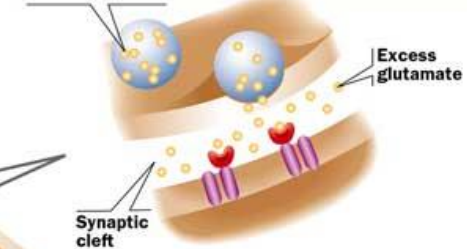
A nerve impulse is transmitted when the terminal fibers of one neuron's axon release chemicals called neurotransmitters that attach to dendrites of the receptor neurons.



## 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.

### Neurotransmitter molecules



Dulcie Teesateskie/Huntsville Times

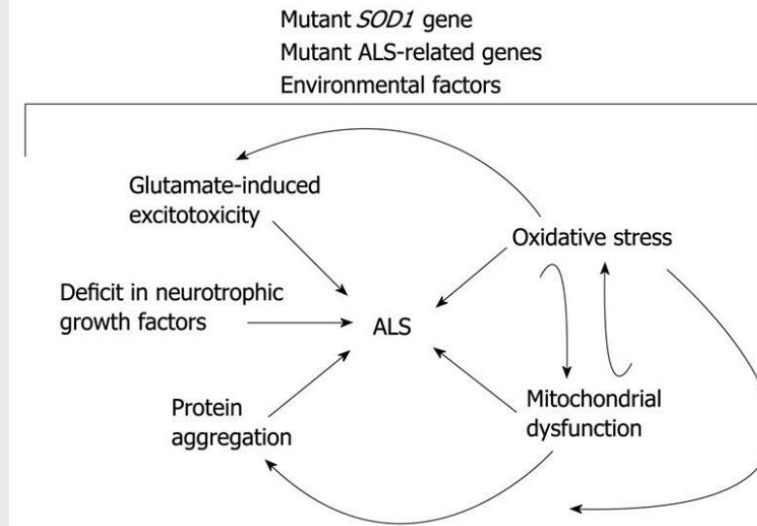
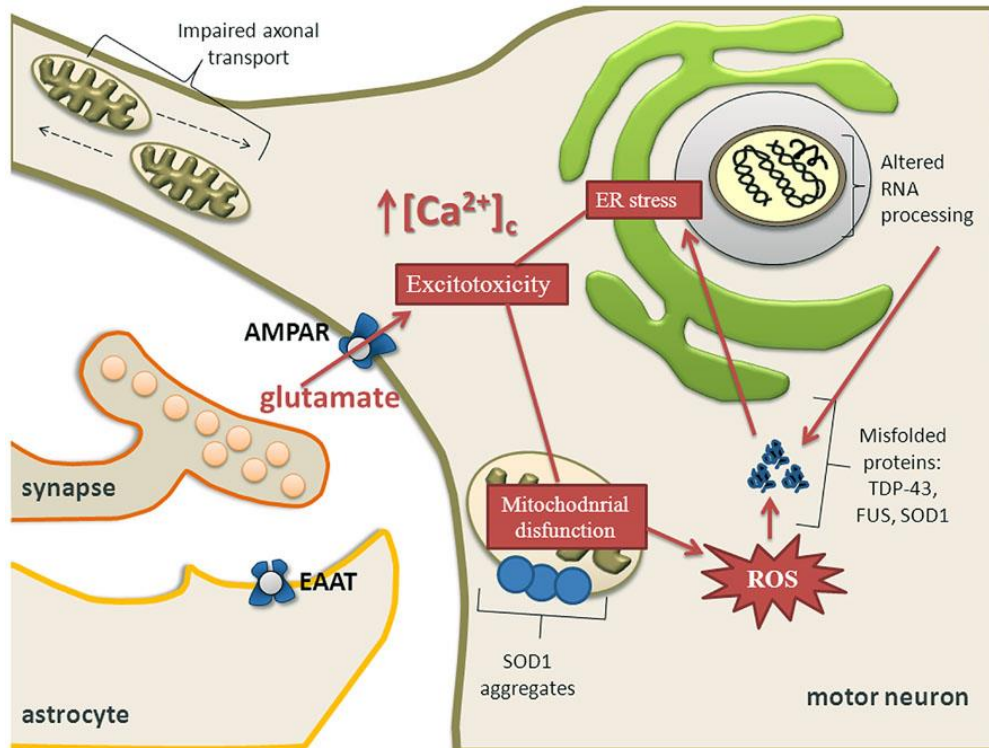
# Amyotrophic Lateral Sclerosis

- ▶ Amyotrophic lateral sclerosis (ALS) is the most common motor neuron disease in adults
- ▶ The incidence of ALS is approximately **1–2.6 cases per 100 000 persons annually**, whereas the prevalence is approximately **6 cases per 100 000**.
- ▶ Those who were in the Gulf War are more likely to get ALS compared with other veterans.
- ▶ 20% more common in men than in women
- ▶ Typically midlife, between 40-70 years- most common Caucasian
- ▶ Life Expectancy is 2-5 years, more than ½ live longer than 3 years

On June 1, 2021, a team of scientists led by the [NIH](#) and the Uniformed Services University announced it had discovered a unique form of genetic ALS that affects children as early as age 4 years. This childhood form of ALS is linked to the gene ***SPTLC1***

# Etiology

- ▶ Sporadic , unkown cause
- ▶ Genetic in 5-10% of cases-  
an inherited autosomal trait



# Etiology

## ▶ Theoretical Models

- ▶ Excitotoxicity- Glutamate defect in possibly metabolism, transport or storage
- ▶ Oxidative stress caused by free radicals
- ▶ Autoimmune-antibodies to calcium channels, activated T lymphocytes , Monoclonal paraproteinemia
- ▶ Cytoskeletal defects or abnormalities
- ▶ Neurofilament abnormalities- abnormal accumulation and damage to the structure

# 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

# Treatment for ALS

▶ **The U.S. Food and Drug Administration (FDA) has approved drugs to treat ALS:**

- **Riluzole** (Rilutek) is an oral medication believed to reduce damage to motor neurons by decreasing levels of glutamate, which transports messages between nerve cells and motor neurons. Clinical trials in people with ALS showed that riluzole prolongs survival by a few months, particularly in the bulbar form of the disease. Individuals with swallowing difficulties may prefer the thickened liquid form (Tiglutik) or the tablet (Exservan) that dissolves on the tongue.
- **Edaravone** (Radicava) is given by intravenous infusion and has been shown to slow the decline in clinical assessment of daily functioning in persons with ALS.



medications to help manage symptoms of ALS, including muscle cramps, stiffness, excess saliva and phlegm, and the 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, and constipation.

# 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