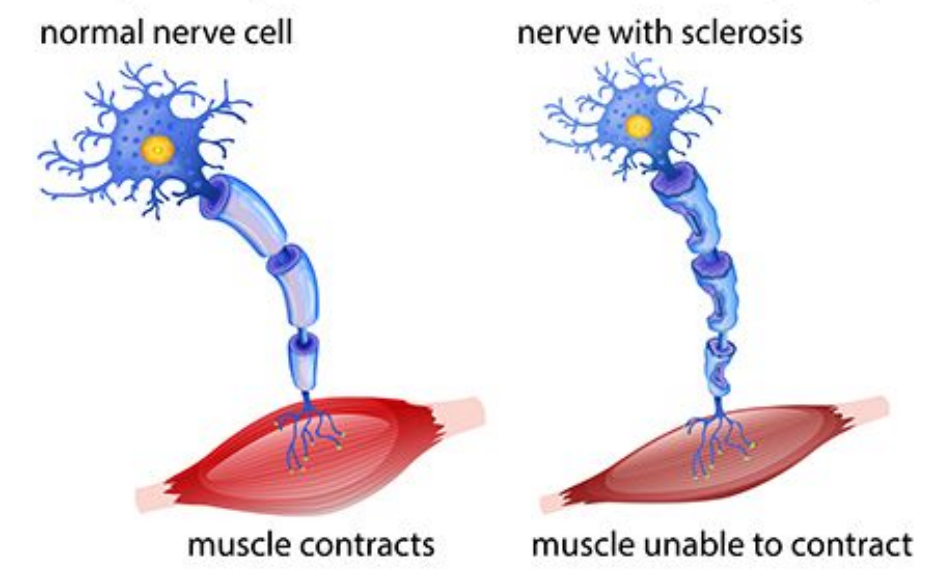

**Note: Click and hover over the audio file on each slide to play voice over content.

Amyotrophic Lateral Sclerosis

Emmaline Hill, Angelica Goliat, Jessica Lopez, Marilou Mendoza, Julian Ramirez, Caitlin Hume

“Amyotrophic” comes from the Greek language. "A" means no. "Myo" refers to muscle. "Trophic" means nourishment. So, amyotrophic means "no muscle nourishment,” and when a muscle has no nourishment, it "atrophies" or wastes away.

"Lateral" identifies the areas in a person's spinal cord where portions of the nerve cells that signal and control the muscles are located.

Pathophysiology

Also known as Lou Gehrig's disease, is a neurodegenerative disorder that causes progressive muscle weakness and wasting, leading to paralysis of respiratory muscles.

The **hallmark feature** of ALS is the selective degeneration and death of motor neurons in the brainstem, spinal cord, and motor cortex. These motor neurons are responsible for transmitting signals from the brain to the muscles, allowing for voluntary movement of the limbs, trunk, and face.

Is progressive and no established treatment, which is 100% fatal.

Etiology/causes are widely unknown

- 5-10% of cases = Inherited by genetic mutation
  - This form is known as “Familial”
- 90-95% of cases = No known cause with no family history
  - This form is known as “Sporadic”
  - Most common in US and occurs randomly
  - Likely contributing risk factors include environmental, lifestyle

- Researchers continue to study possible causes of ALS
- **Most theories center on a complex interaction between genetic and environmental factors**

Symptoms

Symptoms are progressive and may affect differently depending on location of motor neuron degeneration.

Early

- Soft weaker or tight and spastic muscles
- Twitching
- Fatigue
- Poor balance
- Dysphagia
- Dysarthria

Symptoms

Middle

- Up to 90% paralysis
- Worsened Dysphagia
- Worsened Dysarthria
- Disoriented speech
- Difficulty breathing due to compromised associated muscles of the lungs
- Constant headache, dizziness
- Disturbed emotions: Uncontrollable laughter or crying
- Anxiety, depression

Late

- Paralysis
- Disability to use lung muscles
- Cardiac arrest
- Choking

Risk Factors

Risk Factors

- Family inheritance
- Genetics
  - C9OF72, SOD1, TARDBP genes
- Age
  - 40s - mid 60s
- Sex
  - Males > Females
- Environmental factors
  - Smoking, Toxin exposure
  - Occupation

Diagnostics

Diagnostic Tests

- Brain and spinal cord MRI
- Muscle nerve biopsy
- Lab tests - Including CBC, UA, thyroid function
- Lumbar puncture to test CSF
- Electromyography (EMG) and nerve conduction study (NCS) to evaluate muscle and motor neurons

Potential Complications

Potential Complications:

- Inability to produce purposeful movement → pressure sores, DVT, PE
- Dysphagia → malnutrition, dehydration, weight loss
- Aspiration → pneumonia
- Loss of communication → depression and anxiety
- Loss of respiratory control → respiratory failure & ventilator dependence

NCLEX Qs

#1: A client with ALS is admitted to the hospital for management of symptoms. Place the following interventions in the correct order of priority for the nurse to implement:

1. Administer prescribed medications to manage pain and discomfort.
2. Teach the client and family about the use of mobility aids to maintain independence.
3. Assess the client's respiratory status and administer oxygen therapy if needed.
4. Evaluate the client's ability to swallow and provide interventions as necessary.

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Correct order: 3, 4, 1, 2

ALS experience progressive muscle weakness, including the muscles used for breathing and swallowing. Respiratory status should be assessed first and oxygen therapy provided if needed, as respiratory failure is the leading cause of death in ALS. Swallowing ability should be evaluated and interventions provided as necessary to prevent aspiration and malnutrition. Pain management is an important aspect of care for clients with ALS, but it is a lower priority than addressing respiratory and swallowing issues. Finally, mobility aids should be addressed to help the client maintain independence, but this is a lower priority than addressing urgent physiological needs.

SOURCE: Jackson, M. J., & Swami, N. (2020). Nursing management of amyotrophic lateral sclerosis. Nursing Made Incredibly Easy!, 18(6), 33-40.

#2: Which diagnostic test can be used to diagnosis of ALS?

A. Electromyogram
B. Muscle Biopsy
C. Serum Creatinine Kinase
D. Cerebrospinal Fluid Analysis
E. Pulmonary Function Test

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Abnormal cerebrospinal fluid findings include elevated proteins, and an increased white blood cell count. CSF electrophoresis reveals an increase in the myelin basic protein and the presence of increased immunoglobulins, especially IgG.

Ignatavicius: Medical Surgical Nursing (p. 874)

#3: The client is being evaluated to rule out ALS. Which key features would the nurse note to support the diagnosis?

A. Loss of consciousness
B. Muscle weakness and spasticity
C. Seizure activity
D. Cardiac dysrhythmias

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ALS results from the degeneration and demyelination of motor neurons in the spinal cord, which results in weakness and spasticity of the muscles.

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Thanks!

Image courtesy of mymed.com illustrating ALS.