Mitsubishi Tanabe Pharma America

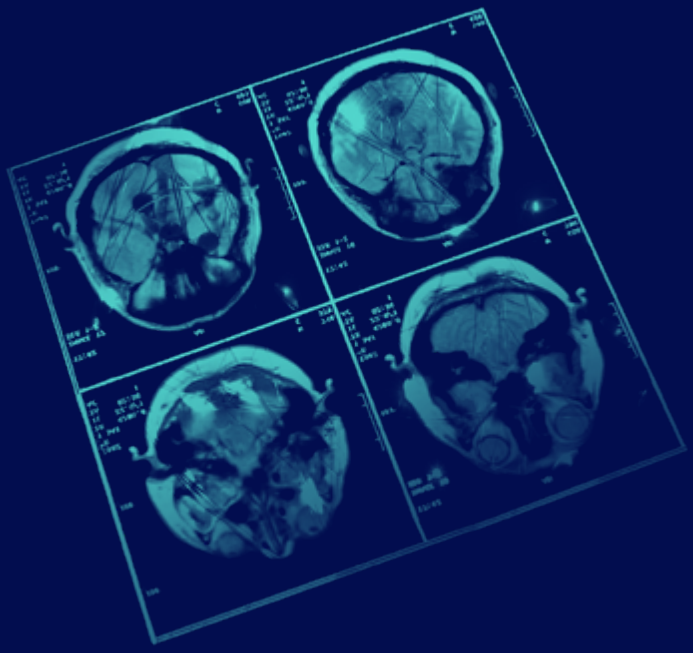
# Understanding Amyotrophic Lateral Sclerosis (ALS)

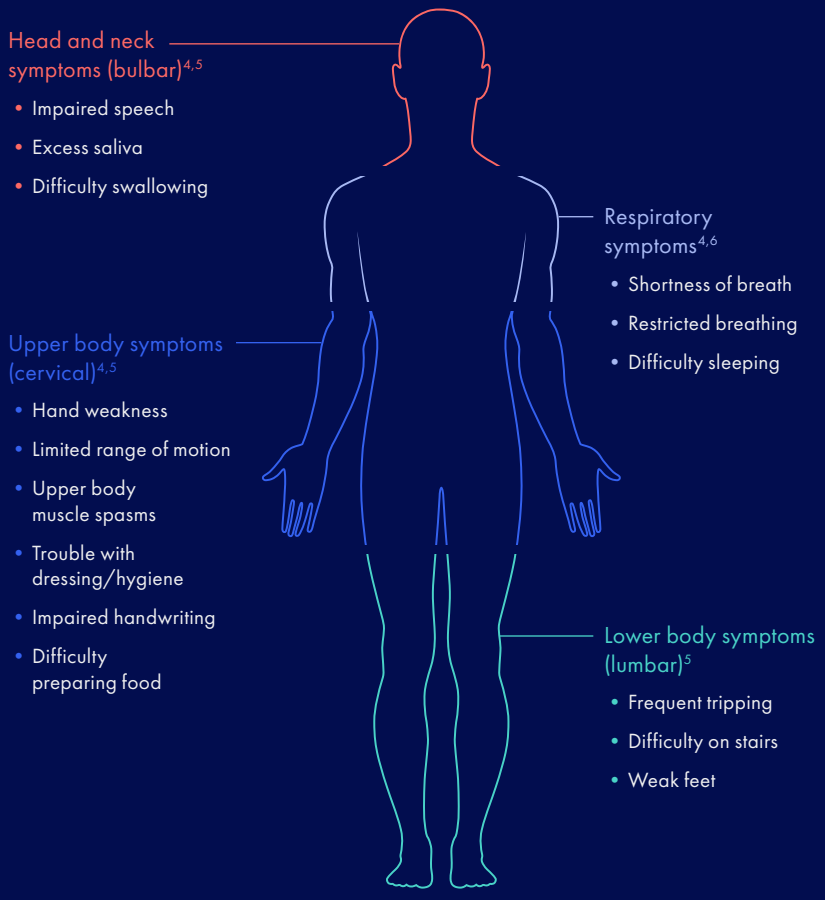
## What is ALS?

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a nervous system disease that impacts physical function.[1] ALS begins in the brain and spinal cord by affecting nerve cells called motor neurons, which control voluntary muscle movement such as chewing, walking, breathing and talking.[1]

In people with ALS, these motor neurons stop working, resulting in the brain's inability to communicate with the muscles and causing them to become weak and lose functionality.[1]

ALS is a progressive disease, meaning it involves a gradual onset of symptoms.[1,2] People with ALS lose their strength and muscle function over time, which can limit their ability to live independent lives.[1] The average life expectancy after diagnosis is two to five years, but progression of the disease can vary significantly.[3]






## Symptoms & Diagnosis

Symptoms vary from person to person and can appear in different regions of the body.[1] The number and degree of symptoms typically increase as the disease gets worse.[1]

No one test can provide a definitive diagnosis of ALS.[7] ALS is primarily diagnosed based on medical history and symptoms observed by a physician along with a series of tests to rule out other mimicking diseases.[7] However, the presence of upper and lower motor neuron symptoms is a strong indicator of ALS. Initial symptoms can be subtle at first, and it can take 12 to 14 months to be accurately diagnosed with ALS.[8]

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# Who gets ALS?

ALS is the most common of the motor neuron diseases (MNDs), which is a wider group of disorders that can lead to loss of physical function.[1] It affects people of all ages, races and ethnic backgrounds.[1]

An estimated 5,000 people in the United States (U.S.) are diagnosed with ALS each year.[9]

On average, a new case of ALS is diagnosed every 90 minutes.[9]

**5-10%**
of ALS cases are familial, meaning they are due to genetic mutations and are inherited from a family member.[7]

**90-95%**
of ALS cases are sporadic, meaning the cause(s) of the disease are unknown.[7]

Although ALS can strike at any age, symptoms most commonly develop between the ages of 40 and 70, and average age at diagnosis is 55.[7]

For unknown reasons, military veterans are more likely to be diagnosed with the disease than the general public.[10]

# Treatment & Monitoring

There is no cure for ALS; however, there are treatments available that can help control symptoms, help prevent or delay complications and slow the progression of the disease.[8] This includes medications approved by the U.S. Food and Drug Administration (FDA), physical and speech therapy, respiratory support and nutritional support.[8]

Several clinical tools that help monitor ALS symptoms and track disease progression have been developed, including:

**Function-based questionnaires** that ask people with ALS to rate how well different muscle groups are working based on a sliding scale, providing a high-level assessment of overall muscle function. The ALS Functional Rating Scale-Revised (ALSFRS-R) is a validated rating instrument for monitoring disability progression in ALS and widely used in clinical trials.[11]

**Strength measurements** to assess muscle weakness and help physicians understand how quickly ALS is progressing. The most common method for this is called handheld dynamometry (HHD), in which the examiner holds a small gauge that the patient pushes against using different muscle groups.[11]

**Respiratory measurements** to assess the possibility of respiratory dysfunction, the leading cause of death among people with ALS. The most typical test for this is a forced vital capacity (FVC) test, which measures the maximum amount of air a patient can exhale from their lungs after taking the deepest possible breath.[1]


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