Amyotrophic Lateral Sclerosis (ALS)

Amyotrophic lateral sclerosis (ALS), often referred to as “Lou Gehrig’s Disease” (named after a famous baseball player who had the disease) is a rare neurologic disorder characterized by degeneration of motor neurons in the brainstem and spinal cord. It causes progressive muscle weakness leading to death. Patients will require respiratory support and palliative care at the end of life.



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Assessment

Progressive Muscle Weakness
Progressively Weaker Muscle
Muscle weakness begins distally especially in the hands of most patients and progressively affects all muscles including the diaphragm. This disease has no cure and the muscle weakness progresses causing dysarthria and dysphagia, difficulty speaking and difficulty swallowing due to motor muscle weakness. It is important to assess aspiration and choking risk in these patients and modify communication methods as needed.

Systemic Muscle Wasting
Systemic Muscle Waste-basket
As the muscles weaken, they atrophy and become smaller. Patients experience full body muscle wasting. In later stages of the disease, patients are bedridden.

Fasciculations
Fast-pickle
Small muscle twitches are common especially in the face, because neurons degenerate and misfire.

Spasticity
Space-tick
Loss of upper motor neuron function in the brain leads to a disconnection with the body’s lower motor neurons, which results in spasticity. Daily moderate exercise and physical therapy assist with maintenance of ADLs and mobility.

Fatigue
Sleepy-guy
Systemic muscle weakness often leads to fatigue or a general feeling of exhaustion. Exercising with a focus on endurance can decrease fatigue in many patients.

Considerations

Riluzole (Rilutek)
Reel-u
While this disease has no cure, Rilutek (riluzole) is the treatment of choice. It works by reducing glutamate excitotoxicity, mainly through inhibition of glutamate release and inactivation of voltage-gated sodium channels. Other medications like phenylbutyrate-taurursodiol and edaravone are also FDA-approved and have been shown to slow the rate of functional decline.

Stretching
Stretching
Moderate daily exercise increases muscle endurance and decreases spasticity. Daily exercise of moderate intensity is an important treatment to prolong survival.

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Little to No Cognitive Decline

Little Down-arrow Brain to No Down-arrow Brain Signs

While this disease progressively weakens muscles, patients usually have no decrease in cognitive abilities.

Respiratory Support

Lungs being Supported

As the disease progresses, patients will need long-term respiratory support, as the diaphragm is weakened. Initially patients require C-PAP or Bi-pap only during sleep but progress to ventilator dependent states. Patients most commonly expire due to respiratory infections.

Palliative Care

Palliative-pail

Palliative care is a type of medical care that focuses on relief from symptoms, pain, and stress of the condition for those with serious, chronic, and life-threatening illnesses. The goal is to provide support and improve quality of life for the patient and family.

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