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A Brief Note on Amyotrophic Lateral Sclerosis

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Editorial

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ABOUT THE STUDY

Amyotrophic Lateral Sclerosis (ALS), also known as Motor Neuron Disease (MND) or Lou Gehrig's disease, is a neurodegenerative disease that results in the progressive loss of motor neurons that control voluntary muscles. ALS is the most common type of motor neuron disease. Early symptoms of ALS include muscle stiffness, twitching, and gradually increasing weakness and muscle wasting. Limb-onset ALS causes weakness in the arms or legs, whereas bulbar-onset ALS causes difficulty speaking or swallowing. Half of people with ALS have at least mild cognitive and behavioral problems, and about 15% develop front temporal dementia. The vast majority of people are in pain. Chewing, speaking, and walking are all controlled by the affected muscles [1]. Motor neuron degeneration progresses to the point where the ability to eat, speak, move, and, eventually, breathe is lost.

Since at least 1824, Charles Bell has described the disease. Jean-Martin Charcot, a French neurologist, first described the link between the symptoms and the underlying neurological problems in 1869, and coined the term amyotrophic lateral sclerosis in 1874. Motor neurons that are affected by ALS can be classified. In typical or "classical" ALS, upper motor neurons in the brain and lower motor neurons in the spinal cord are involved [2].

Primary Lateral Sclerosis (PLS) affects only upper motor neurons, whereas Progressive Muscular Atrophy affects lower motor neurons (PMA). There is some debate over whether PLS and PMA are separate diseases or simply variants of ALS. Regional ALS symptoms are limited to a single spinal cord region for at least a year; they progress more slowly than classic ALS and have a higher survival rate [3].

Cognitive or behavioral dysfunction affects 30-50% of ALS patients. Around half of people with ALS will experience mild cognitive and behavioral changes, and 10-15% will develop Front Temporal Dementia (FTD). The majority of people with ALS who have normal cognition at diagnosis have preserved cognition throughout their disease; the development of cognitive impairment in those with normal cognition at baseline is associated with a poor prognosis [4]. However, in people with ALS, cognitive and behavioral dysfunctions have been found to correlate with decreased survival and increased caregiver burden; this may be due in part to deficits in social cognition. Over time, people's ability to move, swallow (dysphagia), speak, or form words deteriorates (dysarthria). Upper motor neuron involvement is characterized by tight and stiff muscles (spasticity) and exaggerated reflexes (hyperreflexia), including an overactive gag reflex [5]. An abnormal reflex known as Babinski's sign also indicates upper motor neuron damage. Symptoms of lower motor neuron degeneration include muscle weakness and atrophy, cramps, and fleeting twitches of muscles visible beneath the skin (fasciculation) [6]. Twitching, on the other hand, is a side effect rather than a diagnostic symptom; it occurs after or in addition to weakness and atrophy. The death of both upper and lower motor neurons (located in the motor cortex of the brain) is the defining feature of ALS (located in the brainstem and spinal cord). In ALS with front temporal dementia, neurons in the frontal and temporal lobes of the brain die [7]. The presence of Bunina bodies, which are inclusion bodies (abnormal protein aggregations) in the cytoplasm of motor neurons, is a pathological hallmark of ALS. Aside from motor neuron death, the majority of ALS variants share two other characteristics: focal initial pathology, which means symptoms begin in a single spinal cord region, and progressive continuous spread, which means symptoms spread to additional regions over time.

Mutations in TARDBP and FUS are thought to increase the low-complexity domain's binding affinity, causing the proteins to aggregate in the cytoplasm [9]. Once these mutant RNA-binding proteins have misfolded and aggregated, they may be able to misfold normal protein within and between cells in a prion-like manner. This also leads to lower levels of RNA-binding protein in the nucleus, which could indicate that their target RNA transcripts are not being processed normally. The physician's observations of the patient's symptoms and signs, as well as a battery of tests to rule out other diseases, are used to make the diagnosis of ALS [9]. Physicians obtain the patient's complete medical history and usually perform a neurologic examination at regular intervals to determine whether symptoms such as muscle weakness, atrophy, hyper reflexia, and spasticity are worsening. A number of biomarkers are being researched for the condition, but none are currently in widespread medical use [10]. Although there is no known cure for ALS, the drug Riluzole has been approved for treatment and may slow progression of the disease. It is expensive, however, and appears modestly effective. Generally, treatment is designed to help control symptoms. Drugs such as baclofen or diazepam may help control spasticity.

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