Amyotrophic Lateral Sclerosis

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Amyotrophic Lateral Sclerosis (ALS)

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Pathophysiology

- Reactive astrocytes display increased immunoreactivity for the proteins GFAP and S100B. They express inflammatory markers, like COX-2, neuronal NOX (Saberi et al., 2015).
- Microglia are activated in response to neuronal distress and release proinflammatory cytokines and reactive oxygen species as a result (Saberi et al., 2015).
- Ubiquitin, a dense, round structure found in the cytoplasm of neurons in the anterior horn cells, is indicative of ALS (Saberi et al., 2015).
- PICT-1 is a translocator protein found in muscles fibres called NOGO-A (Gordon, 2013).
- Formation of pathological protein aggregates, like ubiquitin, is in the cytoplasm of the cells (Saberi et al., 2015).
- Animal research is inhibited by a pathological protein found in muscles fibres called NOGO-A (Gordon, 2013).

Pathophysiology Continued

- TDP-43 is a ribonucleic protein that stabilizes mRNA, and assists with its processing, transport, and translation (Saberi et al., 2015).
- In those with ALS, there is a loss of function in TDP-43 and related formation of pathological aggregates, like ubiquitin, in the cytoplasm of the cells (Saberi et al., 2015).
- The morphology of cell death in ALS is not clearly understood, but research points to cell apoptosis (Saberi et al., 2015).
- Axonal regrowth is inhibited by a pathological protein found in muscles fibres called NOGO-A (Gordon, 2013).

Familial ALS

- Supernovae mutations in SOD1 mutations accounts for 20% of familial ALS (ALS). (Saberi et al., 2015)
- FUS gene has younger average age for onset, predominantly starts in the legs with lower motor neuron disturbances, and has low occurrence of cognitive disturbances (Gordon, 2013).
- TDP-43 is the most frequently occurring gene mutation in ALS, accounting for up to 40% of cases (Gordon, 2013).
- TDP-43 accumulation in the brain and deposits of p62 show up processes like ubiquitination and p62 (Gordon, 2013).

Significance of Pathophysiology

Because ALS affects upper and lower motor neurons functionally, eventually it affects the entire body. The affected person starts noticing minor help with activities of daily living (ADLs), but eventually, loss of motor neuron function impacts body movements to the point that the affected person is more dependent on others for all ADLs.

The most common complication leading to death is respiratory failure. Some affected by the disease are chosen to initiate life prolonging measures such as placement of a PEG or intubation and ventilation, but others may refuse supportive care outright. Since there is no cure for ALS, and only one drug available on the market that has been shown to slow disease progression minimally, getting multiple supportive services into the home early has been proven to provide great benefit to the affected person and their loved ones through greater quality of life and quantity of life. (Williams et al., 2013)

Implications in Nursing

The implications of care for someone with a life limiting and incurable disease shift the locus to supportive measures such as diet, respiratory therapy, home medical equipment, physical therapy, or counseling.

Ensuring that nursing care focuses on comfort, aligns with the patient and family’s goals of care, and employs a multidisciplinary approach is key. Nurses need to use their resources and make referrals as appropriate. Nurses will also need to be vigilant in assessing for skin breakdowns for the bedbound and nutritionally impaired, assessing for respiratory infections and encouraging good pulmonary habits, and guiding patients and family’s on food choices and safe swallowing techniques as dysphagia presents itself.

Communication in later stages of ALS can become problematic for many patients. Coming up with creative ways to communicate and encouraging early discussions about what a person wants for their future care will help guide families and clinicians alike through the terminal stages of the disease.

References

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