Kindred at Home

Physician to Physician:

Hospice & Palliative Management of Amyotrophic Lateral Sclerosis (ALS)

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Amyotrophic Lateral Sclerosis (ALS) is an incurable neurodegenerative disorder affecting both upper and lower motor neurons resulting in motor weakness, disability, and eventually death. First described by Charcot in the 19th century, ALS became better known as Lou Gehrig's disease when the acclaimed New York Yankee major league baseball player and seven-time All-American suddenly retired at the peak of his career because of ALS. Gehrig died two years later in 1941. Later generations identify ALS with renowned theoretical physicist and cosmologist Stephen Hawking, who was born less than a year after Gehrig's death. Hawking was diagnosed with ALS in 1963 and is remarkable, among many things, for living with ALS for over 50 years until his death in 2018.

Even with today's improved supportive measures, the median life expectancy with ALS, from symptom onset until death, is only three to five years, though as many as 10% of individuals with ALS may live up to ten years. The motor neuron effects of ALS can impact any muscle group, eventually leading to neuromuscular respiratory failure, dysphagia with nutritional impairments, and/or aspiration, with these being the ultimate cause of death. Most patients with ALS also experience other non-motor neurologic findings, ranging from autonomic dysfunction such as constipation to sensory symptoms including paresthesias or neuropathic pain. Other types of pain are experienced in up to 85% of patients with ALS due to immobility, muscle cramps, muscle spasms, or other musculoskeletal causes.

This broad range of findings is best managed by a multidisciplinary team, as is found at tertiary ALS centers, which is associated with improved survival and quality of life. Palliative care can provide additional support, further improving quality of life throughout the course of the illness. When an individual's prognosis is six months or less, hospice care provides comparable interdisciplinary care with greater emphasis on comfort and preparation for death. Hospice is especially helpful as patients become less able to access outpatient settings and express a desire to not return to the hospital. Establishing goals of care with advance care planning should be accomplished early in the course of ALS, with these revisited periodically as the disease progresses. Enhancing communication between patients and families with attention to psychosocial, spiritual, and practical support are hallmarks of best practice, alongside impeccable palliative management of pain, dyspnea, and other symptoms. Hospice excels at accomplishing these needs, emphasizing care of not only the patient, but also the involved family, even after death with bereavement services.

Many variables influence prognosis in ALS, including decisions to pursue or forgo life-sustaining treatments such as non-invasive mechanical ventilation (NIMV) or medically assisted nutrition and hydration (MANH). Some individuals with ALS choose not to pursue NIMV or MANH, viewing these as prolonging the dying process. Others opt for these with their associated life extension. When a patient chooses to forgo such measures at the point in time that they are offered, this is often the indicator that the patient is likely within six months of their expected lifespan, meaning hospice referral is indicated. When life-sustaining measures have been instituted, ascertaining the appropriate time for hospice referral can be more complex. Hospice professionals can assist in making such determinations, using other signs, such as recurring infections despite antibiotics, progressive nutritional decline despite MANH, development of non-healing pressure wounds, or if the individual expresses a desire to withdraw NIMV or MANH.

Patients with ALS benefit significantly from the additional care provided by hospice. Unfortunately, many hospice referrals involving ALS are very late, within days or weeks of death. While that may seem appropriate if the referral is based on a decision to withdraw life-sustaining measures, it is important to understand why the patient is making that decision, such as increasing symptom burden or failure to maintain nutrition despite enteral feeding. Involving hospice at the first sign of such changes is often appropriate, with hospice being able to manage ALS patients even while they continue so-called 'aggressive measures' like NIMV or MANH, as long as the likely prognosis is six months or less. Therefore, it is often better to introduce hospice early on. Even if prognosis is greater than six months, hospice professionals can educate the patient and family regarding options with hospice admission at some future time. Waiting to refer until one is sure that hospice is needed often results in days, weeks, or even months of needless suffering. Please consider contacting hospice sooner, rather than later, to avoid 'brink-of-death' hospice admissions that fail to provide all the support that hospice can deliver.

Let us know how we can assist with care for your patients with ALS. We are here.